MENTAL RETARDATION
ABSTRACTS

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Rehabilitation Services Administration
Division of Developmental Disabilities
Washington, D.C. 20201

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Ms. Mary Herner
Project Director
Herner Information Services, Inc.
2100 M Street, N.W.
Washington, D.C. 20037

Ms. Patricia Thoben
Project Officer
Division of Developmental Disabilities
Rehabilitation Services Administration, SRS
Washington, D.C. 20201

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Psychological Bulletin
Psychological Monographs
Psychological Record
Psychological Reports
Psychological Review
Rehabilitation Counseling Bulletin
Rehabilitation Literature
Science
Training School Bulletin

BROAD ASPECTS OF MENTAL RETARDATION

2441 **MacKAY, D. N.** Prevalence of mental subnormality in Northern Ireland and North-East Scotland. *Lancet,* 1(7713): 1357, 1971. (Letter)

Prevalency surveys of low (IQ 0-19) and medium (IQ 20-49) grade MR in the geographically and demographically similar areas of Northern Ireland and Northeast Scotland reveal that higher rates for both grades of MR in nearly every age grouping occur regularly in the Northern Ireland province. This finding is in line with previous studies which have reported a similarly higher rate of incidence of mongolism when the 2 areas are compared. (5 refs.) - *N. Mize.*

Muckamore Abbey
Co. Antrim, Northern Ireland

2442 **ARNOLD, IRENE L.; & *BUDNER, STANLEY.** Research for service to severely retarded children. *Children,* 18(3): 101-104, 1971.

The primary focus of Retarded Infants Services (RIS) in New York City is service to severely retarded children and their parents, but it also undertakes research to discover the best methods of providing service with the limited financial resources available to a relatively neglected area of the population, the severely retarded child. Several studies which have been conducted and which show concrete results or policy recommendations are reported. The significance of the RIS research experience stems from the fact that viable research programs may be conducted by small agencies with limited financial resources, in order to use the feedback results to develop and enlarge present agency information and referral services to clients. (No refs.) - *T. C. Turner.*

*Fordham University
New York, New York

2443 **CHASEY, WILLIAM C., JR.** Exposure to handicapped children as a variable in the self-perception of college undergraduate students. *Therapeutic Recreation Journal,* 5(1): 22-24, 1971.

Ss were 30 college students who volunteered to serve as student clinicians and 25 undergraduate students who constituted the control group. Student clinicians worked with the same handicapped child for an 8-week session on gymnasium activities; controls had no contact with the children. The Fiedler Interpersonal Perception Scale for the measurement of self-concept was given to the 30 clinicians and 25 undergraduates 1 week before the clinic began and at the end of the 8-week clinic. The criterion variable employed was the difference between pretest and posttest scores. Twenty-two (73%) of the clinicians showed a more favorable perception of self at the completion of the session, while no significant change in the self-perception of the control group was evident. Student clinicians tend to have a more positive view of self-perception after working with handicapped children. (7 refs.) - *T. C. Turner.*

University of Texas at Austin
Austin, Texas

2444 **TOPPEN, J. T.** Underemployment: Economic or psychological? *Psychological Reports,* 28(1): 111-122, 1971.

The Wechsler Adult Intelligence Scale and 4 tests for measuring psychopathic tendencies were given to 50 able-bodied chronically underemployed men to assess their individual psychological characteristics in order to discover possible causes for their chronic underemployment. Tests were given orally. A control group of 50 Ss was also tested. The underemployed group's scores did not parallel normative levels as much as did the control

group; there was more psychopathy among the underemployed, who, percentagewise, were significantly further outside the healthy and normal range than the control group. IQ scores for the unemployed group were also substantially lower, the highest percentage (40%) of them falling within the 70-79 WAIS range, as compared to the highest percentage (about 35%) of the control group within the 90-99 range. Even after interaction of uncontrolled variables of race and education were statistically excluded, greater incidence of aberrant test scores among the underemployed continued. Since serious psychological aberration of one or another type was found among the 50 underemployed Ss, psychological knowledge of the adverse effects of these aberrations on behavior suggests that the psychological handicap and the underemployment are probably related, perhaps developing early in life and causing a lifelong handicap. (11 refs.) - *T. C. Turner.*

University of Cincinnati
Cincinnati, Ohio

2445 **ELWOOD, J. H.; & PEMBERTON, J.** Infant mortality in Belfast. *Archives of Disease in Childhood,* 46(247): 336, 1971.

The infant mortality rate for Belfast for 1963-1965 was 29.4/1,000 live births. This was significantly ($p < 0.001$) higher than the rate for Birmingham for 1964, which was 21.4/1,000 live births. Differences between these distributions by parity accounted for only 12% of the excess. However, differences by birthweight accounted for 40% of the excess infant mortality in Belfast. There was a much higher rate of deaths attributed to postnatal asphyxia and atelectasis in Belfast than in Birmingham; 6.6/1,000 and 0.4/1,000, respectively. The Belfast rate was also much higher than the rate for England and Wales. Rates for congenital malformations were 5.7/1,000 in Belfast and 4.9/1,000 in Birmingham. (23 refs.) - *J. K. Wyatt.*

Institute of Clinical Science
Belfast 12, Ireland

2446 **MOTULSKY, ARNO G.** Human and medical genetics: A scientific discipline and an expanding horizon. The William Allan Memorial Award Lecture. *American Journal of Human Genetics,* 23(2): 107-123, 1971.

Scientists working in human and medical genetics need to take stock of the current status of the field and to propose a plan for future effort. High priority areas include research on population, reproductive genetics, aggression, effects of environmental agents and their interaction with specific genotypes, and the relationship of specific genotypes to the environment. Unlike many other scientific fields, the one of human and medical genetics usually does not have independent disciplinary status with a consequent slowness of institutional growth and a lack of departments with strong graduate training programs, even though medical geneticists are needed in many specialty areas of medicine. The field has, however, a strong laboratory basis, and recent findings are leading to still more new discoveries. While considerable knowledge has been gained in areas such as biochemical genetics and cytogenetics, poorly developed areas include those of behavioral genetics, pharmacogenetics, and population, reproductive, and developmental genetics. Much of the work in genetics impinges on areas of high public interest and considerable public interaction occurs in genetic counseling, genetic engineering, and the genetic components of many common diseases. Appropriate development of adequate resources to provide for early implementation of findings for public use is an important responsibility in this field. (No refs.) - *M. S. Fish.*

University of Washington
Seattle, Washington 98105

2447 **SAPPENFIELD, BERT R.** Abnormal orientations: A revision of nomenclature. *Psychological Reports,* 28(2): 456-458, 1971.

The medical model of psychiatric nomenclature has been criticized for treating all deviations from normal perception, thinking, and behavior as disorders, illnesses, or diseases. According to a more appropriate psychological model, the term "orientation" is a useful substitute for such terms as "disorder," "illness," "disease," "disturbance," and "reaction." "Orientation" applies to all forms of normal functioning as well as to abnormal forms. With use of this term, therefore, it is easy to communicate that psychological

principles extend across the whole range of human orientations and that no new principles are needed to account for abnormal orientations. (10 refs.) - *B. J. Grylack.*

University of Montana
Missoula, Montana

2448 **PAYNE, JAMES S.** Prevalence survey of severely mentally retarded in Wyandotte County, Kansas. *American Institute for Mental Studies Training School Bulletin*, 67(4): 220-227, 1971.

A survey of SMRs in Wyandotte County, Kansas, showed a higher incidence rate than that found in other prevalence studies. Criteria for inclusion in the study were IQ of 50 or below (mongols below 5 without IQ scores were also included), previous diagnosis by a psychologist, county residence, family interview, and living at the time of the survey. The survey, which covered 482 families, operated in 3 stages: Agency correspondence, solicitation of names from Agency, and personal contacts. Data on race revealed that the county was 83.2 percent White and 16.8 percent nonwhite; MRs were 68.5 percent White, 31.3 percent nonwhite, and .2 percent unknown. Twenty mothers and 28 fathers of MRs had less than a sixth-grade education; 13 mothers and 17 fathers were college graduates. The youngest mother of an MR child was 15 years old; the oldest was 49. The youngest father was 16; the oldest was 68. Visual, hearing, speech, and orthopedic impairments were reported among the MR children. (16 refs.) - *B. Berman.*

University of Virginia
Charlottesville, Virginia

2449 State of Illinois Department of Mental Health. *Statistical Classification of Mental Disorders and Mental Retardation.* Springfield, Illinois, 1970, 33 p.

This set of classifications for mental disorders and MR is divided into 11 parts: MR, organic brain syndromes, psychoses not attributable to conditions listed previously, neuroses, personality disorders, psychophysiologic disorders, special symptoms, transient situational disturbances, childhood and adolescent behavior disorders, nonspecific conditions, and nondiagnostic terms. It also is provided with a cross-index. The classification scheme used is that of *Diagnostic and Statistical Manual of Mental Disorders*, second edition. (5 refs.) - *T. C. Turner.*

Department of Mental Health
Springfield, Illinois 62706

2450 **PLESS, IVAN B.; & ROGHMANN, KLAUS J.** Chronic illness and its consequences: Observations based on three epidemiologic surveys. *Journal of Pediatrics*, 79(3): 351-359, 1971.

Epidemiologic surveys relating to the psychological and social consequences of chronic physical disorders were conducted in Great Britain, the Isle of Wight (England), and Monroe County, N.Y. Frequency of chronic illness in the 0-16 years age range varied from 6-11%, compared to 5-20% in other studies (depending on broadness of the definition of physical disorder used). In each of the 3 studies, the major cause of illness was asthma and other allergic disorders. Sensory disorders and nervous system disorders, especially epilepsy and cerebral palsy, ranked second and third. At least 1/3 and probably more of the chronic illnesses of childhood were likely to be permanent. Twelve to 13% of the disorders were severe (requiring prolonged hospitalization or confinement to bed), 37-53% were mild (condition interfered only with strenuous activities such as athletics), and the remainder were of moderate severity. The only demographic characteristic differentiating the chronically ill from healthy controls was the higher ratio of boys among the chronically ill. Secondary handicaps in the areas of educational achievement, behavior, and psychologic maladjustment were more frequent among the 111 Ss than among healthy children. Up to 30% of the ill Ss may be handicapped by secondary social and psychologic malfunctioning, with possibly 1/2 of such malfunctioning attributable to the physical disorders. More effective comprehensive services are need to prevent secondary handicaps. (12 refs.) - *K. L. Woodworth.*

Rochester Child Health Study
Rochester, New York 14619

2451 **MACKAY, D. N.** Mental subnormality in Northern Ireland. *Journal of Mental Deficiency Research*, 15(1): 12-19, 1971.

In a survey of the prevalence of MR in Northern Ireland, there were 6,117 ascertained MRs in a general population of 1,502,000, indicating a prevalence rate of 4.07/1,000. The MR population consisted of 3,253 males and 2,864 females, and 22% were under age 14 years. Prevalence rates for low IQ (0-19) and medium IQ (20-49) MRs were 0.36/1,000 and 2.43/1,000, respectively. The prevalence rate for Down's syndrome was 0.64/1,000. About 1/3 of the MRs were institutionalized. Inter-regional comparisons suggest that these overall prevalence rates are not reliable indices of prevalence. (16 refs.) - *J. K. Wyatt.*

Muckamore Abbey Hospital
County Antrim, Northern Ireland

2452 BENDEL, R. Methodological problems with ecological variables. *Journal of Mental Deficiency Research,* 15(3): 196-206, 1971.

Wolfensberger and Halliday (1970) (W&H) obtained county data for 10 socio-ecological variables and performed a multiple regression analysis with the institutionalization of retardates as the dependent variable. They obtained a multiple correlation coeffecient of R 0.67, and according to their study, the socio-ecological approach appears promising because family and clinical variables can account for no more than 50 percent of the variance. Bendel discusses some of the methodological problems involved with the W&H data, showing that neither of the above statements are warranted. The independent variable must be removed from the regression before inferences can be made about the factors affecting rate of institutionalization. R then drops to 0.34, showing that W&H data can account for only 12 percent of variance. Alternative "stepwise" techniques give much different results with the W&H data, especially the backward elimination and "best" method of stepwise regression (best subset of predictors which significantly explain the variation). As a result, the socio-ecological approach as shown by Wolfensberger and Halliday's data does not appear to be a promising area in MR research. (12 refs.) - *T. C. Turner.*

Pacific State Hospital
Pomona, California 91768

2453 FLETCHER, JOSEPH. Ethical aspects of genetic controls: Designed genetic changes in man. *New England Journal of Medicine,* 285(14): 776-783, 1971.

The ethical question of justification of genetic changes in man for the sake of therapeutic and non-therapeutic benefits is discussed. In this problem both the means and ends are challenged or condemned. Fletcher suggests that the rock-bottom issue and definitive question in the ethical analysis of genetic control is whether to reason from general propositions to normative decisions or from empirical data to normative decisions. To the priori moralist, good consequences do not justify bad acts or means. The principle of proportionate good would not, then, make genetic intervention by laboratory reproduction morally permissible. The more commonly held ethical approach is a pragmatic one, in which one reasons from the data of each actual case and then chooses the course offering maximum or optimum desirable consequences. On that basis the ethical issue is whether genetic change in man will add or detract from human welfare. When would the utility of constructive eugenics justify it? Some situations are suggested: store tissues of endangered human groups for later replication; specialize the capacities of people by cloning. The danger of tyranny is real, but genetic controls do not lead to dictatorship in a cause-and-effect manner; the problem of possible misuse is political, not biological. Fletcher also ethically defends genetic research in the laboratory and comments that most discussion regarding ethics of biomedical intervention is a semantic mess, since there is no overall consensus on what we mean by "human" or "humanistic". The historic moral order has always presupposed heterosexual coital conception as necessary for the continuance of life and this is no longer the case. As new means of biological intervention arise, the necessity for multiple social decisions of profound consequence is coming. (17 refs.) - *T. C. Turner.*

Box 358, University Hospital
Charlottesville, Virginia

2454 DAVIS, BERNARD D. Ethical and technical aspects of genetic intervention. *New England Journal of Medicine,* 285(14): 799-801, 1971. (Editorial)

Techniques and moral aspects of gene therapy are considered. There is fear that the techniques used to cure hereditary diseases by genetic manipulation could also be used to control human behavior. With such new problems, and in a time of moral flux, solutions will come only through extensive debate. Two opinions are noted: Paul Ramsey's conservative view and Joseph Fletcher's pragmatic view, emphasizing the relevance of risk-benefit calculations. No act is good/bad in and of itself; moral judgment of any act should be based on its consequences—a gene is not good/bad in the abstract, but its value depends on how it functions within the genetic context and environment of the individual. Knowledge will continue to be amassed about genetics and rather than run away from the linked threat and promise of discovery, society must try to learn how to increase the ratio of benefits to costs. There are already ways to diagnose hereditary diseases and prevent them by abortion. Cure of a hereditary disease by gene transfer of a diploid nucleus is much easier and already available in amphibia and is what most needs to be publicly discussed now. The required moral guidelines cannot, however, be derived from medicine; the question is rather, when, how far, and at what risk will mankind decide to apply to itself its growing knowledge of genetics and evolution. (4 refs.) - *T. C. Turner.*

2455 KESTELMAN, PHILIP. Mortality of abortion. *Lancet,* 2(7724): 607, 1971. (Letter)

Aborting and sterilizing a woman are more dangerous than continuing her pregnancy, and considerably more dangerous than simply aborting her, especially during the first trimester. In England and Wales, in 1969, there was only 1 death in 29,864 abortions alone, compared with 12 in 11,126 abortions plus sterilization. There are, further, various alternatives to female sterilization: vasectomy of the husband and contraception (barrier, intrauterine, oral). Buckley-Sharp's Chi-square test has greatly clarified the statistical associations between mortality and abortion. (12 refs.) - *B. Berman.*

10 Carston Close
London, SE12 8DZ England

2456 SELVIN, STEVE; & GARFINKEL, JOSEPH. Estimation of the rate of congenital malformations. *Pediatrics,* 48(3): 492-493, 1971.

An alternative way of perceiving the estimation of the rate of congenital malformation from multiple sources has been reported. When death certificates are matched to birth certificates, 3 possible outcomes exist with respect to congenital malformations: the malformation will be noted on both certificates, it will be included on the birth certificate only, or it will be noted on the death certificate alone. These 3 outcomes can be considered as a fourfold table with 1 cell missing, namely the cell in which a malformation is present but not noted on either the birth or death certificate. Equations based upon the proposed tables were used to present certain new data on the rates of congenital malformation in New York State during the years 1959 to 1967. (2 refs.) - *B. J. Grylack.*

Birth Defects Institute, 84 Holland Avenue
Albany, New York 12208

2457 PLESS, IVAN B.; & SATTERWHITE, BETTY. Health education literature for parents of handicapped children. *American Journal of Diseases of Children,* 122(3): 206-211, 1971.

There is a generally acknowledged need for educating parents of handicapped children. If guilt feelings arise from a misunderstanding of the nature of the condition and the extent to which it was preventable, they may lead to inappropriate child-rearing patterns and to secondary psychological or social handicaps. The pediatrician is responsible for conveying accurate and complete information regarding a child's illness, but may not always be skillful in communicating with parents. Written materials are therefore strongly recommended as an aid. The authors assembled a list of booklets and pamphlets (Table 1: Health Education booklets; Table 2: Biographical Literature) to use in disseminating information to the parents of the afflicted. Written materials alone cannot adequately meet parents' need for information; the physician must spend time in discussing the child's diagnosis. Some of the problems in communication may be attributable to the confusion inherent in trying to provide comprehensive health care of the chronically ill child in a health system which is organized primarily toward episodic care. If secondary psychological and social consequences

are to be avoided, health literature can be one pillar upon which communication between parent and physician can be built. (18 refs.) - *T. C. Turner.*

University of Rochester
School of Medicine and Dentistry
Rochester, New York 14620

2458 **RUBINSHTEIN, S. IA.** L. S. Vygotsky on the psychological development of mentally retarded children. *Soviet Education*, 14(1,2,3): 6-19, 1971-1972.

The uniqueness of the mental development of MR children has been the subject of exhaustive analysis by Soviet psychologists, L. W. Vygotsky among them. In assessing the psychological development of the MR child, Vygotsky proposes the differentiation of nuclear signs of MR from secondary and tertiary stratifications. According to his theory, the underdevelopment of higher mental functions is the initial and most frequent complication to occur as a secondary symptom of MR, yet it is not necessarily related to MR itself. Higher forms of memory, thinking, and character are the product of cultural development rather than of biological maturity. In the mentally handicapped child, cultural development is fully possible but is limited because of such nuclear signs of MR as inadequate perception of everything new and insufficient degree of activity. In the light of these concepts, it appears to be necessary to reorganize the system for rearing the MR child in infancy and preschool years. This theory has prompted the search for more effective methods of raising and training MRs and has emphasized the importance of verifying nuclear signs of MR. (7 refs.) - *B. J. Grylack.*

Moscow Institute of Psychiatry
RSFSR Ministry of Health
Moscow, U.S.S.R.

2459 **MENOLASCINO, FRANK J., ed.** *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 386 p.

This book summarizes contemporary psychiatric viewpoints and treatment-management techniques in the areas of education and behavioral management of MR. The individual articles are grouped under the headings of diagnostic issues and methods, treatment and management approaches, family dimensions, the prevention of MR, and current psychiatric training programs in MR. The presentations are of all types and generally admonish the psychiatric profession to participate more fully in all aspects of MR. A further aim is to interpret present psychiatric views and involvement in MR for all other professionals concerned with MR. - *S. Glinsky.*

CONTENTS: Mental Retardation: the Cinderella of Psychiatry (Potter); Problems of Emotional Development in Young Retarded Children (Webster); Psychopathology and Mental Retardation (Philips); Autism in Children with Mental Deficiency (Bender); Psychosis of Infancy and Early Childhood as Manifested By Children With Atypical Behavior (Reiser); The Facade of Mental Retardation: its Challenge to Psychiatry (Menolascino); Mourning and the Birth of a Defective Child (Solnit & Stark); Mental Retardation and Family Stress (Begab); What Do You Tell the Parents of a Retarded Child? (Solomons); Emotional Conflicts of Handicapped Young Adults and Their Mothers (Mowatt); Parents of the Mentally Retarded: an Operational Approach to Diagnosis and Management (Menolascino); Treatment of the Emotional Problems of the Retarded Child and of the Family (Chess); Psychotherapy of the Mentally Retarded: Values and Cautions (Lott); Psychiatric Programs for Very Young Retarded Children (Woodward, Jaffe & Brown); Head-banging: its Meaning and Management in the Severely Retarded Adult (Collins); Nurturant Nursing of Mentally Retarded Ruminators (Wright & Menolascino); Psychiatric Aspects of Mongolism (Menolascino); Panacea, Palliation or Poison: the Psychodynamics of a Controversy (Kurlander & Colodny); Drug Effects on Learning in Children: a Selective Review of the Past Thirty Years (Freeman); Pregnancy Stress and Maternal Perceptions of Infant Endowment (Cohen); Prevention of Autism in a Young Infant in a Well-Child Clinic (Call); Mental Subnormality and Child Psychiatry (Tizard); Development of a Training Program in Mental Retardation for Psychiatric and Pediatric Residents (Cytryn & Milowe); Mental Retardation and Comprehensive Training in Psychiatry (Menolascino); General Principles of Training Programs for the Mentally Retarded (Collins).

2460 **PHILIPS, IRVING.** Psychopathology and mental retardation. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 39-47.

The MR is particularly vulnerable to maladaptive behavior and MR nomenclature is outmoded. Diagnostic studies including psychiatric evaluation were made of 227 families with an MR child between 1958 and 1965 at Langley Porter Neuropsychiatric Institute. The Ss had all degrees of MR, were preponderantly brain-damaged, and ranged in age between 9 months and adolescence. The vulnerability of the MR derives both from constitutional endowment as well as from interpersonal experiences with his environment. Common misconceptions regarding MR maladaptive behavior include assignment of such behavior to MR rather than interpersonal relationships, the belief that MR emotional disorders are different in nature from those in normals, and that certain symptomatologies derive from organic brain damage. Lastly, outdated MR concepts and nomenclature should conform to the present state of knowledge and be discarded for more functional classifications. (13 refs.) - *S. Glinsky.*

University of California School of Medicine
San Francisco, California

2461 **TIZARD, JACK.** Mental subnormality and child psychiatry. In: Menolascino, Frank, J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 329-347.

Advances have been made in mental subnormality and similar progress is possible elsewhere in child psychiatry. Although the mentally subnormal form the largest class of severely disordered children provided for under the Mental Health Services, MR has been relatively neglected by child psychologists and psychiatrists working in other fields as well as by pediatricians, educators and psychiatric social workers. At present a great deal is known about the epidemiology, causes and treatment of mental defect. Much less is certain about the etiology of MR. Systematic long-term studies hold the key to progress in these areas. (26 refs.) - *S. Glinsky.*

University of London Institute of Education
London, England

MEDICAL ASPECTS – Diagnosis (General)

2462 **SCHWARTZ, ROBERT P.; & *PARKE, JAMES C., JR.** Rapid screening test for protein and glucose in cerebrospinal fluid. *Journal of Pediatrics*, 78(4): 677-680, 1971.

A "urine dipstick" has been found useful in the detection of abnormal concentrations of cerebrospinal fluid (CSF) protein and glucose. CSF was examined for a group of 43 patients, ages 1 day to 12 years, with findings suggestive of CNS disease. These included: fever of unknown origin, septicemia, meningitis, seizure disorders, CNS leukemia, microcephaly, dehydration, hypoglycemia, and ataxia. Results from 58 specimens analyzed by immersing the dipstick (Labstex brand of reagent strips) in the CSF and comparing to quantitative laboratory examinations gave correlations which showed that, although the dipstick readings provided only rough approximations, the approach has value in obtaining quick and useful screening of CSF for glucose and protein when quantitative determinations are delayed. Protein values above 40 mg% and glucose values below 50 mg% were readily detected. (7 refs.) - *M. S. Fish.*

*Charlotte Memorial Hospital
Charlotte, North Carolina 28201

2463 **FLUHARTY, ARVAN L.; LASSILA, ELTON L.; PORTER, MYNA T.; & KIHARA, HAYATO.** The electrophoretic separation of human galactosidases on cellulose acetate. *Biochemical Medicine*, 5(2): 158-164, 1971.

An improved procedure for the fractionation of human β-galactosides by cellulose acetate electrophoresis is described. The technique utilizes more routinely available equipment (a Gelman Delux Chamber with a Beckman Duostat power supply, Sepraphore III cellulose polyacetate strips, and a Chromato-Vue cabinet) than does the starch-gel procedure, is faster, and requires smaller amounts of sample. Examination of the effects of voltage, time, pH, and type and strength of buffer on the migration and resolution of enzymatic components has delineated optimum conditions for performing the assay. The system is adaptable for the determination of β-glucosidase and β-N-acetylglucosaminidase. Of particular interest may be the increased sensitivity which can facilitate the examination of β-galactosidases in extracts of tissue culture cells derived from Type I, II, and III patients with mucopolysaccharidoses. (15 refs.) - *M. S. Fish.*

Pacific State Hospital
Pomona, California 91768

2464 **GOTOFF, SAMUEL P.; GADZALA, CECILE; YING, ROSITA L.; & WENDELL, PAMELA W.** Relationship of neonatal IgM values to congenital abnormalities and mental retardation. *Journal of Pediatrics*, 78(6): 1020-1025, 1971.

The use of routine screening of cord serum IgM levels to detect subclinical intrauterine infection does not appear to be justified. Quantitative determination of IgM concentrations in 4,285 samples of sera obtained from 3 separate hospitals resulted in the identification of 68 (1.6%) sera with elevated IgM values (>17 mg percent). A total of 1,659 samples from one hospital which serves a predominantly low-income, Negro population had 41 (2.5%) sera with elevated values, compared to another hospital serving Caucasian, middle-class communities, which provided 1,635 samples, 23 (1.4%) of which were elevated. The study group, consisting of the infants with elevated IgM values, had the same total number of neonatal complications (68) as did the control group. Examination of members of these groups at 40 weeks consisted of a developmental screening inventory, derived from the Gesell Developmental Schedules, and performed for 53 study and 54 control infants. Of these latter groups, the study and control Ss were normal in 45 and 48 of the cases, respectively. Two major developmental abnormalities were observed in each of

these 2 groups. Ten percent of the study group had neonatal complications, but these were recognized clinically without the use of IgM levels. The majority of the infants with congenital anomalies had normal cord blood IgM values. While the results of this study and of other previous reports do not support the use of routine screening of cord serum for IgM, the difference in the instances of elevated values observed for the two types of populations is significant. (20 refs.) - *M. S. Fish.*

Abraham Lincoln School of Medicine
Department of Pediatrics
Chicago, Illinois 60612

2465 PAUL, RICHARD H.; & HON, EDWARD H. Clinical fetal monitoring. A survey of current usage. *Obstetrics and Gynecology*, 37(5): 779-784, 1971.

An evaluation study has been carried out to assess the use of direct clinical fetal monitoring techniques as a potentially valuable clinical tool. Of a total of 67 obstetric units known to use fetal monitoring to which questionnaires were sent, 61 responded to the original and one follow-up questionnaire, and 55 of these provided adequate response to most areas covered. Besides general information related to the number of deliveries per year, the number monitored, and the length of time monitoring had been used, the evaluation included the use of the FECG electrode, intrauterine catheter function, and fetal heart rate (FHR) patterns. Inquiries related to how these monitoring techniques were utilized, problems and complications associated with their use, and the interpretation of results. In the use of the FECG electrode 28 services applied the device in the labor bed, 4 in the delivery room, and 23 in both places. No serious complications with this technique were reported. The intrauterine catheter was utilized in 33 institutions, and only 9 of these experienced moderate to marked difficulty with the insertion; however, more difficulties with catheter insertion than with FECG electrode application were reported. Thirty-six institutions identified and recognized FHR patterns with no difficulty, and 41 stated that FHR patterns aided in the ability to predict the condition of the newborn. (27 refs.) - *M. S. Fish.*

LAC/USC Medical Center
Los Angeles, California 90033

2466 SYMONDS, E. MALCOLM; WILLIAMS, STEPHANIE S.; & CELLIER, KEVYN M. Maternal and fetal influences on the acid-base balance of human amniotic fluid. *Obstetrics and Gynecology*, 37(5): 742-749, 1971.

A study of factors involved in the control of amniotic fluid acid-base balance during labor has shown that a significant relationship exists between fetal and amniotic fluid carbon dioxide tension, standard bicarbonate and amniotic fluid pCO_2 and standard bicarbonate over a wide range of normal and abnormal acid-base values. From a group of 104 women in labor, selected because of signs of fetal distress or a maternal complication, amniotic fluid samples were obtained from 56 Ss during labor and 27 Ss at the time of delivery. The acid-base balance of these samples was compared with those obtained at the same time from fetal and maternal blood. In addition to the above findings, study of the clinical conditions of the infants at birth disclosed that pCO_2 of amniotic fluid at delivery was significantly lower in depressed than in healthy infants. Failure of transfer of CO_2 may be caused by fetal circulatory collapse associated with severe asphyxia. Due to the loss of CO_2 the pH of amniotic fluid alters rapidly on exposure to air, although standard bicarbonate remains constant. (12 refs.) - *M. S. Fish.*

Queen Victoria Hospital
Adelaide, South Australia

2467 A new way to watch over newborns. *Medical World News*, 12(16): 20E, 1971.

CABAS—Computerized Automated Blood Analysis System—was developed by a University of Utah team to monitor blood gases in newborns. The system draws blood samples through 2 catheters in the umbilical artery, which permits establishment of cerebral arterial oxygen concentrations. With CABAS it is possible to determine whether an infant's physical status is outside the range the physician thinks would give the baby a chance to survive, and then to institute vigorous therapy where necessary. Careful preliminary evaluation attempts to assure that the infant's condition will not be adversely affected by the monitoring procedure. First used on puppies, the system has been shown to be quite effective, but it has not yet been approved for routine diagnosis. (No refs.) - *B. Berman.*

2468 **POLANI, NINA.** Dermatoglyphic topology. *Developmental Medicine and Child Neurology*, 13(2): 235-238, 1971.

Topology may be applied to palmar dermatoglyphics in order to add this dimension to the analysis of chromosomal aberrations. Frequencies of configurations have been developed based on a description of all palmar loops and a count of all triradii. Both autosomal and sex chromosomal anomalies show variable loop densities in specific areas. The ratio of loops to triradii appears to be greater with an additional Y chromosome and less with an additional X. (20 refs.) - *E. L. Rowan.*

Guy's Hospital Medical School
London S.E.1, England

2469 **NELSON, GEORGE H.; WAGES, HARVEY S.; HAHN, DOROTHY A.; AZIZ, EZZAT M.; & FREEDMAN, DON S.** A complication of fetal scalp blood sampling. *American Journal of Obstetrics and Gynecology*, 110(5): 737-738, 1971.

The breaking off of the exposed part of the blade in the fetal scalp at the time of the stab wound occurred in an infant subjected to fetal scalp blood sampling. The infant was delivered approximately 3 hours following the procedure and had a 1-minute Apgar score of 7. One piece of metal was removed, but it was elected to leave the remaining piece of blade in place. At 4 months of age, the infant was developing normally, and the metal tip was evident in the same location. Investigation revealed that the exposed tip of the blade had broken off from the piece of metal embedded in the Teflon Sheath. (4 refs.) - *B. J. Grylack.*

Medical College of Georgia
Augusta, Georgia

2470 **MACGILLIVARY, IAN.** Ultrasound in the measurement of fetal growth. *Developmental Medicine and Child Neurology*, 13(1): 106-107, 1971.

Although ultrasound measurements are useful in determining whether fetal growth is occurring at the normal rate, they are not always accurate in measuring actual maturity, and, since certain hazards may be associated with their frequent use, they should be limited in number. A review of a group of studies has shown that unanimous agreement does not exist for how well ultrasound measurements can determine maturity. Since several investigators utilized various different parameters in these studies (different populations of women and various types of complicated pregnancies), a true assessment of the technique is difficult. A need exists for measurements which are representative of a normal pregnant population, are carried out serially in women of known gestation length, and are correlated with the percentile weight of the baby and the degree of maturity at delivery. Hazards may include the possible deleterious effect of ultrasound on chromosomes; as a consequence, scans should be limited in number, particularly during early pregnancy; at this time they have little value, since determination of biparietal diameter is unsatisfactory before 16 weeks. (10 refs.) - *M. S. Fish.*

University of Aberdeen
Foresterhill, Aberdeen AB9 2ZD, Scotland

2471 **MOORE, W. M. O.; MURPHY, P. J.; & DAVIS, J. A.** Creatinine content of amniotic fluid in cases of retarded fetal growth. *American Journal of Obstetrics and Gynecology*, 110(7): 908-910, 1971.

Specimens of amniotic fluid and serum were obtained from 21 mothers during the last trimester of pregnancy in order to measure the creatinine content of amniotic fluid and to determine its reliability as an index of gestational age in normal and retarded fetal growth. Amniocentesis was performed in cases of suspected isoimmunization where bilirubin estimation was required and cases of suspected intrauterine growth retardation where liquor volume and cytology studies were required. Amniotic fluid creatinine was found to increase with advancing gestation and appears to be a reliable index of gestational age where fetal growth is normal. Amniotic fluid creatinine concentration is not a reliable index of maturity if the fetus is small for gestational age, however, because low values occur. While low creatinine values are indicative of prematurity when fetal growth is normal, such values may be associated with maturity when fetal growth is retarded. (9 refs.) - *B. J. Grylack.*

St. Mary's Hospital
Manchester M13 OJH, England

2472 **MILLER, MAURICE H.** Neonatal and infant auditory screening programs. *Clinical Pediatrics*, 10(6): 340-345, 1971.

Detection of hearing defects should begin in the neonatal period and should be done with audiometric testing devices. Current practice of observing responses to a 3000 Hz stimulus presented at a level of 90 dB SPL gives many false positives and negatives and inadequate information on normal maturation, fails to detect hereditary and unilateral impairments, and is negated by "auditory recruitment" (abnormal increase in loudness of a sound as its intensity increases above the impaired threshold), which interferes with detection of some significant hearing impairments. Proper screening tests can detect conditions that interfere with normal speech and language maturation, such as MR, cerebral auditory problems, minimal cerebral dysfunction, and emotional disturbance. Greatest need is detecting the child with an auditory defect or other communication disorder during the first year of life (conductive hearing loss associated with serious otitis media is frequent in newborns). The 3000 Hz stimulus, at the present developmental stage, should not be used in mass screening. (16 refs.) - *B. Berman.*

New York University Medical Center
New York, New York

2473 **JOHNSON, CHARLES FELZEN; & OPITZ, ERICA.** The single palmar crease and its clinical significance in a child development clinic. *Clinical Pediatrics*, 10(7): 392-403, 1971.

Palmar crease studies were made of 276 children referred to a Child Development Clinic for problems of delayed development or other anomalies, and of 150 well children who served as controls. A single palmar crease (SPC) is present when the distal and proximal horizontal creases are replaced by or joined into a single crease. Approximation of the 2 lines with bridging was read as an incomplete SPC. Of the 276 Ss, 18 (6.5%) had a SPC (9 unilaterally, 9 bilaterally) and 13 (4.7%) had an incomplete SPC (11 unilaterally, 2 bilaterally). Among the controls, only 3(2%) had a SPC. Ss with palmar crease abnormalities were found more likely to have borderline or lower intellectual ability($p=<0.05$) than those without palmar abnormalities, and 16(52%) of Ss with crease abnormalities had associated congenital anomalies as compared to only 73(30%) with normal creases. In addition to the SPC, other palmar crease abnormalities were found among 23 of the 276 Ss; 17 of these had associated congenital anomalies, but only 7 had borderline or lower intelligence. Palmar crease abnormalities are seen to suggest some insult to the fetus during the first trimester (when palmar creases are being formed), with the same insult causing the crease abnormality and any associated congenital malformations and lowered intellectual functioning. (15 refs.) - *K. L. Woodworth.*

University of Iowa
Iowa City, Iowa 52240

2474 Is your pediatrician doing his homework? *Expectations*, 1(2): 1, 1971.

Some pediatricians feel that children with learning disabilities do not constitute a strictly medical problem and are therefore neglecting to become well informed enough to treat the child as a medical problem or to do further research in the area. A checklist of questions is given by which to view one's pediatrician: 1) Has he shown knowledgeable interest in learning disabilities and discussed the problem? 2) Has he recommended a neurological examination and professional tests for visual and motor perception? 3) Has he given the child a complete physical, including a 5 hour glucose tolerance test and basal metabolism, allergy, vitamin, and mineral deficiency tests? 4) Has he considered medication for hyperactivity, or a nutritious diet, vitamin supplements, and exercise? 5) Does he work closely with the school? 6) Does he keep abreast of current research, realizing that not all children outgrow their learning disabilities? If the answer is not "yes" on these points, then the pediatrician may not believe that a child's learning disability is a medical problem. (No refs.) - *T. C. Turner.*

2475 **WARBURG, METTE.** Norrie's disease. *Eye, Birth Defects*, 7(3): 117-124, 1971.

Thirty-six cases (6 families) in Sweden and Denmark have revealed Norrie's disease, a male affliction caused by a particularly pleiotrophic gene on the X chromosome and manifesting opthalmologic, psychiatric, and otologic signs in a progression rendering unrecognizable the initial

malformations. At birth or in the initial months of life the disease shows bilateral blindness, with a white retrolental membrane, and the eyes usually phthisic. Retinal dysplasia with typical malformative rosettes may be seen. In 9 cases, severe progressive MR occurred between ages 18 months and 5 years; these stopped walking and talking alone, became disturbed, and lost control with others. Eleven adult blind males had impaired hearing. Dementia or psychosis appeared in one-fourth of the cases. The most important differential diagnosis is bilateral retinoblastoma, and a correct diagnosis is mandatory for survival. In some cases, retrolental fibroplasia of premature babies subjected to oxygen therapy, as well as toxoplasmosis, may present problems. Other differential diagnoses include falciform detachment, juvenile retinoschisis, congenital retinal detachment, and sex-linked microphthalmia and cataract. (18 refs.) - *B. Berman.*

National Institute for the Blind
Copenhagen, Denmark

2476 **VEASY, GEORGE L.; CLARK, JUSTIN S.; JUNG, A. LARRY; & JENKINS, JARRELL L.** A system for computerized automated blood gas analysis: Its use in newborn infants with respiratory distress. *Pediatrics,* 48(1): 5-17, 1971.

Computerized automated blood analysis system (CABAS) is a method which has been devised for computerized on line, real time analysis of arterial Pao_2, $Paco_2$, and pH for use in newborn infants with respiratory distress. The blood gas measurements are based upon a 0.3 ml blood sample withdrawn automatically from an indwelling arterial catheter, results being displayed subsequently in α-numeric terms on a cribside memory scope. Laboratory studies of 51 dogs and limited clinical trials with 6 infants suffering from respiratory distress suggest that the CABAS is a safe system for blood gas analysis in ill neonates. As a stand alone system, the CABAS would probably cost approximately $20,000. The system may also be of use for other critically ill infants. (13 refs.) - *B. J. Grylack.*

Primary Children's Hospital
Salt Lake City, Utah 84103

2477 **MACHT, JOEL.** Operant measurement of subjective visual acuity in non-verbal children. *Journal of Applied Behavior Analysis,* 4(1): 23-36, 1971.

Five nonverbal retarded children, aged 7 years and younger, and 2 literate adults were used in a study which undertook to develop a procedure for measuring the subjective distance acuity of very young or retarded children. A lever press apparatus was employed to signify a visual discrimination by means of a Snellen "E" discriminanda. The data obtained demonstrated that a subjective measure of distance visual acuity is procurable from nonverbal retarded children. It was apparent from the research that certain critical features must be incorporated into a design geared to obtain information from children. The behavior repertoire of a child must be sufficiently inclusive to afford emission of an objectively observable response. The employed stimulus conditions which eventually control responding should be scheduled adequately. The positive nature of the findings revealed in this study brought into question earlier research which concluded that a subjective method of refraction is of little value in children younger than 7 to 8 years of age. (24 refs.) - *B. J. Grylack.*

University of Denver
Denver, Colorado

2478 **FISHBEIN, MORRIS.** New frontier: Prenatal medicine. *Medical World News,* 12(27): 56, 1971.

The technique of amniocentesis represents significant progress towards medical control of birth defects. Of 370 cases, transabdominal amniocentesis was successful in 353, with no fatalities reported. In this series, abnormalities predicted in 41 fetuses were confirmed in 35 cases, with subsequent termination of these pregnancies. Of the 300 infants who were delivered, all were normal except 2 with Down's syndrome, which had been detected *in utero.* More than 300 genetic disorders in addition to Down's syndrome can be detected with amniocentesis. Risks involved in the technique may include bleeding, inflammation, blood-group sensitization of the mother, and possible damage to or abortion of the fetus. Therefore, specific requirements should be met before intrauterine diagnosis is undertaken. It is recommended that the use of amniocentesis be limited to high-risk groups, such as that in which one parent is a carrier of chromosomal translocations. Although time is needed to

develop the diagnostic possibilities of the technique, within the next 5 or 6 years it may be possible to cure or control certain less severe genetic disorders while the baby is still in the uterus. (No refs.) - *B. J. Grylack.*

No address

2479 **MELANCON, SERGE B.; LEE, SALLY, Y.; & NADLER, HENRY L.** Histidase activity in cultivated human amniotic fluid cells. *Science,* 173(3997): 627-629, 1971.

A study was undertaken to determine if 2 morphologically different cell types (epithelial and fibroblast) had distinct biochemical properties. Histidase (L-histidine ammonia-lyase; E.C. 4.3.1.3) was used as a marker in characterizing cultivated amniotic fluid cells, since histidase activity had been present in epidermal outgrowths of human skin explants in culture, but not in fibroblasts. Amniotic fluid cells obtained at 16 weeks of pregnancy were grown in nutrient medium F 10, containing 11 percent fetal calf serum, penicillin, streptomycin, and fungizone. Fifteen days later, primary cultures were exposed to trypsin for varying lengths of time to separate epithelial and fibroblast cells. Epithelial, fibroblast, and mixed cultures were cultivated in the same medium until the time of histidine assay. The results show relatively lower activity of mixed populations than that found in pure epithelial cells. Fibroblast cultures ranged in activity from none to 1/10 of that in mixed cultures. In only 2 instances were pure fibroblast populations found. This indicates that some culture cells retain tissue-specific properties, as indicated by the presence of histidase activity in epithelial, but not fibroblast, cells. The demonstration of this activity provides a technique for the intrauterine detection of histidinemia, an autosomal recessive disorder characterized by speech defects and occasional MR. The need for establishing the normal biochemical properties for each cell type in amniotic fluid cells is indicated, since this is crucial if the genetic status of the fetus *in utero* is to be detected. (7 refs.) - *T. C. Turner.*

Children's Memorial Hospital
Chicago, Illinois 60614

2480 **TAORI, G. M.; IYER, G. V.; ABRAHAM, J.; & MAMMEN, K. C.** Electrodiagnostic studies in lipidoses, mucopolysaccharidoses, and leukodystrophies. I. Nerve conduction and needle electromyographic studies. *Neurology,* 21(3): 303-306, 1971.

Nerve conduction and needle electromyographic studies were achieved in 1 case of lipidosis, 6 cases of mucopolysaccharidosis, 3 cases of leukodystrophy, and 2 cases of unclassified progressive familial MR. All of the Ss were children. Normal nerve conduction with no fibrillation was found in 4 cases of mucopolysaccharidosis. Normal nerve conduction velocity with fibrillation present was found in the only lipidosis case and the 2 unclassifed cases. Borderline conduction velocity with fibrillation present was seen in 2 mucopolysaccharidosis cases. Severe conduction velocity diminution with fibrillation present was noted in 1 case of leukodystrophy, was absent in the second case, and was not recorded in the third. In conjunction with tools such as urinary enzyme assays, biopsies of nervous and muscular tissue, cerebrospinal studies, and radiographs, nerve conduction and needle electromyographic studies can be of use in diagnosis of problems of familial progressive MR and in differentiation of cases within a given category, such as mucopolysaccharidosis. (15 refs.) - *B. J. Grylack.*

Christian Medical College Hospital
Vellore 4, South India

2481 **KHAZIN, AIDA F.; *HON, EDWARD H.; & YEH, SZE-YA.** Observations on fetal heart rate and fetal biochemistry. III. Base deficit of umbilical cord blood. *Journal of Pediatrics,* 79(3): 406-412, 1971.

Fetal heart rate (FHR) patterns in the 30 minutes immediately preceding delivery were studied in 187 fetuses and correlated with the base deficit values of umbilical artery and vein bloods and with the A-V differences at delivery to determine whether they reflect the status of blood circulation better than the scalp sample does. The mean base deficit of the umbilical artery blood was lowest for normal FHR and increased with the degree of severity of ominous FHR patterns (highest for the FHR pattern of late deceleration). The mean base deficit of umbilical vein blood was also highest for the FHR pattern of late deceleration. Mean umbilical A-V base deficit difference was largest with

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severe and moderate variable deceleration. Findings are similar to those in a previously reported study correlating base deficit of fetal scalp blood samples and FHR patterns. (25 refs.) - *K. L. Woodworth.*

*LAC/USC Medical Center
Los Angeles, California 90033

2482 **SABATER, JUAN; FERRE, CARMEN; & MAYA, ANTONIO.** A misleading finding in urine electrophoresis. *Lancet,* 2(August 7): 316, 1971. (Letter)

Investigation of urinary aminoacids of MR patients revealed 2 cases showing a ninhydrin-positive spot not observed in 15,000 previous chromatographies. The stain was found between glycine and α-alanine, which is an area where no amino acid is normally found. Both patients had been treated with a new drug containing β-hydroxy-γ-aminobutyric acid. When mixed with urine, this substance was found on electrophoresis between glycine and α-alanine. When processed alone, it coincided with histidine. Recognition of this phenomenon is important due to introduction of drugs containing this substance. If methods are used which do not distinguish it from other aminoacids, confusion could arise in evaluating the patient's metabolism. (No refs.) - *K. L. Woodworth.*

Instituto Provincial de Bioquimica Clinica
Barcelona, Spain

2483 **HAYEK, ALBERTO; BAUMAN, ROGER A.; & *CRAWFORD, JOHN D.** ^{99m}Tc pertechnetate for detection of cryptic thyroid tissue in childhood hypothyroidism. *Journal of Pediatrics,* 79(3): 466-468, 1971.

The isotope Tc pertechnetate has been used in over 20 children for a variety of thyroid diagnostic studies with highly satisfactory results. In a 14 1/2-year-old girl with hypothyroidism it was used successfully to detect cryptic thyroid tissue. The S was first seen at 3 years for short stature and started on thyroid treatment after a diagnosis of athyrotic hypothyroidism. No thyroid tissue was felt in the neck or seen at the base of the tongue. At age 14 years the S was restudied. Two weeks after discontinuing thyroid replacement therapy serum thyroxine had fallen to 1.3 μg% and the thyrotropin concentration had risen to greater than 80 mg/ml. One hour after intravenous injection of 1 mCi of ^{99m} Tc pertechnetate, scans revealed localized midline uptake in the sublingual area. Two weeks after the reinstatement of thyroid treatment, serum thyroxine and thyrotropin concentrations had returned to normal. When no goiter is evident, diagnosis of aberrant thyroid tissue requires radionuclides; scanning with ^{131}I.[4] is the conventional procedure. Low uptakes, though, may not permit scanning unless doses of ^{131}I well above levels permissible for diagnostic purposes in children are used. Tc pertechnetate scans are recommended instead to determine presence or absence of thyroid tissue in hypothyroid children when this cannot be determined by physical examination. (5 refs.) - *K. L. Woodworth.*

*Massachusetts General Hospital
Boston, Massachusetts 02114

2484 **GRAY, CLAUDIA; *DAVIDSON, RONALD G.; & COHEN, MAIMON M.** A simplified technique for the culture of amniotic fluid cells. *Journal of Pediatrics,* 79(1): 119-122, 1971.

Forty amniotic fluid specimens ranging from 5 to 100 ml were obtained by transabdominal amniocentesis or at hysterotomy prior to therapeutic abortion with gestational ages varying from 12 to 20 weeks and were used to study techniques for the culture of amniotic fluid cells. In order to ascertain the optimal culture conditions, 3 different treatments were carried out, namely, culture of cells in homologous amniotic fluid, resuspension of cells in various media after centrifugation at 6 x g, and resuspension of cells after centrifugation at 1000 x g. Evaluation according to observation of cell proliferation and ease of obtaining chromosomal preparations indicated that the simplest culture technique apparently yielded the best results, thus showing that the amniotic fluid itself was the best culture medium. Determination of optimal techniques of amniotic fluid cell culture is important for prenatal detection of cytogenetic and metabolic disorders. (8 refs.) - *B. J. Grylack.*

*219 Bryant Street
Buffalo, New York 14222

2485 **BARNET, ANN B.** Evoked potentials in handicapped children. *Developmental Medicine and Child Neurology*, 13(3): 313-320, 1971.

Sensory stimuli evoke cortical electrical potentials which can be separated from background EEG activity by averaging responses to successive stimuli. The auditory evoked response is the best studied and is useful in determining the integrity of peripheral hearing when the subject is unable to cooperate in routine audiometric testing. A great deal of study will be necessary before visual evoked responses can be correlated with localization of visual abnormality. Characteristic evoked responses enable workers to pinpoint gestational age in the neonate and specific abnormalities are noted in some brain-damaged children. A great deal of additional research is necessary but evoked potentials may potentially be of great value in the study of sensory, perceptual, and cognitive function. (45 refs.) - *E. L. Rowan.*

Children's Hospital
Washington, D. C. 20009

2486 **SCHUTT, W. H.** Thumbs in clinical diagnosis. *Developmental Medicine and Child Neurology*, 13(3): 393-394, 1971.

Defects of the thumb are part of many clinical syndromes and may be isolated or part of more general skeletal dysplasia. A rudimentary or absent thumb is part of Fanconi's syndrome and thalidomide embryopathy. Congenital heart disease is also associated with thumb and forearm anomalies. Brachydactyly is part of two multisystem congenital syndromes described by Taybi. The Noack and Carpenter syndromes also involve the thumbs as do Trisomies 18 and 22. Observations of the thumb are helpful not only in clinical diagnosis but also in understanding normal embryogenesis of the extremities. (9 refs.) - *E. L. Rowan.*

Royal Hospital for Sick Children
Bristol BS2 8BJ, England

2487 **PENROSE, L. S.; & LOESCH, DANUTA.** Diagnosis with dermatoglyphic discriminants. *Journal of Mental Deficiency Research*, 15(3): 185-195, 1971.

A comprehensive system of dermatoglyphic diagnosis in certain chromosomal anomalies is explained. A diagnostic discriminant is constructed by weighting the score of each character by the difference between its percentage frequencies in normal and abnormal samples. Following topological principles of classification all loops are recorded as characters and certain triradii are included. The A-line exit can also be included in the discriminant weightings. In examining a given case of chromosomal abnormality for similarity to one or another abnormal type, the dermatoglyphic characters of the patient are scored and multiplied by the discriminative weightings, which belong to each of 4 discriminants (3 trisomic and 1 partially monosomic). Clinical application of the method is demonstrated: (a) in a case of malformation associated with MR; and (b) in examples of mosaicism. The method is useful for screening purposes and weighting calculations could easily be put in the form of a computer program. (17 refs.) - *T. C. Turner.*

Harperbury Hospital
St. Alban's, Hertfordshire, England

2488 **REID, DAVID W. J.; CAMPBELL, DONALD J.; & YAKYMYSHYN, LUELLA Y.** Quantitative amino acids in amniotic fluid and maternal plasma in early and late pregnancy. Preliminary report. *American Journal of Obstetrics and Gynecology*, 111(2): 251-258, 1971.

Twenty-one amino acids were determined quantitatively in amniotic fluid and maternal plasma of 27 normal pregnant women at 2 periods of gestation, and plasma levels of the same amino acids were determined for a group of 16 normal nonpregnant adults in an attempt to establish normal levels of amino acids for 7 to 18 weeks' and 36 to 40 weeks' gestation. At 7 to 18 weeks' gestation, 10 plasma amino acids were significantly lower ($p<0.05$) than corresponding nonpregnancy levels, 6 others were also lower ($p<0.10$), and 5 showed no significant change. At 36 to 40 weeks' gestation, 15 amino acids were lower in amniotic fluid as compared to maternal plasma, and 6 others showed no difference. Sixteen amniotic fluid amino acids decreased during pregnancy, being significantly lower at 36 to 40 weeks' than at 7 to 18 weeks' gestation. These results should provide a basis for

further study of abnormal pregnancies or pregnancies in which mothers are possible carriers of inborn errors of metabolism. (17 refs.) - *B. J. Grylack.*

The University of Alberta Hospital
Edmonton, Alberta, Canada

2489 **SHAYWITZ, BENNETT A.; & RAPIN, ISABELLE.** Hydrocephalus or cerebral palsy—a diagnostic enigma. *Clinical Pediatrics,* 10(9): 546-548, 1971.

Hydrocephalus, one of the most perplexing pediatric neurological entities, in all cases shows defective cerebrospinal fluid absorption in the valve-like mechanisms of the arachnoid villi. However, most clinicians consider it easy to recognize, since standards for comparison and measurement are available. The case of a child is cited who probably showed signs of hydrocephalus as early as 5 or 6 months, but who was not diagnosed until years later. The most important sign is head enlargement, and pediatricians are urged to note divergences from normal head circumference. (6 refs.) - *B. Berman.*

University of Tehran
Tehran, Iran

2490 **SCHIAVONE, D. JOAN; & KALDOR, J.** Serum CPK in brain damage. *New England Journal of Medicine,* 285(13): 753, 1971. (Letter)

Investigation of 22 cases of encephalitis and bacterial meningitis with elevated serum CPK has established that CPK estimation is of definite diagnostic value in CNS inflammation. CPK, apparently of muscle origin, is found also in other toxic cerebral damage, such as overdose and schizophrenia. (No refs.) - *B. Berman.*

University of Melbourne
Heidelberg, Victoria, Australia

2491 **COLES, RUTH A.** Phase contrast microscopy of the placenta. *American Journal of Obstetrics and Gynecology,* 111(3): 369-373, 1971.

Fifty human placentas were studied immediately following delivery in an attempt to determine whether a small piece of placenta obtained with a needle could provide reliable information about the fetus and whether the technique of placental biopsy is safe. Biopsies were obtained from patients who were already anesthetized for cesarean section. The method of placental biopsy was considered unsafe because the aspirating syringe always filled with blood, and it was felt that the chance of causing the placenta to separate was high, particularly if the needle were pressed against a hard infarcted area such as can be found in most placentas. The results of study of the fresh placental material under the phase contrast microscope did not reveal a significant relationship between the state of the fetus and changes in placental villi, and they did not confirm that a few villi are representative of the whole placenta. (12 refs.) - *B. J. Grylack.*

University College Hospital
London, England

2492 **HOBEL, CALVIN J.** Intrapartum clinical assessment of fetal distress. *American Journal of Obstetrics and Gynecology,* 110(3): 336-342, 1971.

Clinical assessment of fetal biochemistry by fetal scalp blood sampling has been helpful in predicting the condition of the fetus at birth. The Ss were 76 fetuses with distress, who were evaluated by continuous fetal heart rate monitoring and fetal scalp blood sampling. Fetuses having moderate to severe heart rate patterns and/or meconium had significant acidosis at some point during labor. Fetuses passing light or heavy meconium were significantly acidotic and hypoxic early in labor, while those with moderate to severe heart rate patterns became acidotic in the active labor phase. Fetuses passing heavy meconium exhibit the greatest morbidity and mortality rates due to meconium aspiration. Those with tachycardia have no biochemical abnormalities during labor, but a high neonatal morbidity. Fetuses showing moderate-severe late deceleration heart rate patterns possess relatively low morbidity and mortality rates, even though they are acidotic during labor and exhibit low one-minute Apgar scores. (16 refs.) - *S. Glinsky.*

Los Angeles School of Medicine
Los Angeles, California

2493 **SIMONSON, E. D.; SCHOEN, JOANNE A.; & BOYD, J. R.** Experiences in assessing fetal acid-base state. *American Journal of Obstetrics and Gynecology*, 107(5): 754-761, 1970.

Saling's technique of fetal scalp sampling has proven in practice to be safe, simple to perform, and an accurate method for evaluating fetal well-being. From September 1966, to February 1969, 159 patients were followed in labor for a total of 234 fetal samplings. Emphasis was placed upon patients with clinical evidence of fetal distress, toxemia, anemia, premature rupture of membranes, and Rh sensitization. Saling's technique of acid-base measurement is far more reliable and accurate than conventional criteria of fetal distress. (13 refs.) - *S. Glinsky.*

Saint Luke's Hospital
Cleveland, Ohio

2494 **CHAN, JAMES C. M.; FRAS, IVAN; WILSON, M. ROBERT, JR.; & MOELLER, HENRY G., JR.** Schizophrenia in infancy, childhood and adolescence. *American Journal of Diseases of Children*, 121(6): 538-542, 1971.

Schizophrenia in infancy, childhood, and adolescence refers to a group of related and overlapping syndromes characterized by withdrawal, regression, and dissociation. Diagnostic criteria include withdrawal from the environment, disturbance of thought and speech, inappropriate affect, alteration in mobility, and disorientation. Further criteria are preoccupation, MR, disintegration of body image, resistance to change, and anxiety. Approximately 1.9% of schizophrenic patients are less than 15 years old. The condition probably derives from a combination of organic and psychogenic factors. Patients respond poorly to treatment, about one-third achieving some social adjustment. No correlation exists between prognosis and therapy used, but the earlier the disorder appears, the worse apparently is the outlook. (49 refs.) - *S. Glinsky.*

Mayo Clinic and Mayo Foundation
Rochester, Minnesota 55901

2495 **KLOPPER, ARNOLD.** Assessment of fetoplacental function by hormone assay. *American Journal of Obstetrics and Gynecology*, 107(5): 807-827, 1970.

Hormone assays are a valuable tool in the clinical evaluation of fetoplacental function. Assays must not be directed to generalized fetoplacental malfunction, but to specific diseases causing specific defects in hormone production. Of the 4 main hormones usually assayed, the role of human chorionic somatomammotropin relative to specific diseases is least clearly understood. Assays of the levels of chorionic gonadotropin, urinary pregnanediol, and estriol are variously useful in abortion, toxemia, retarded fetal growth, diabetic pregnancy, Rh isoimmunization, and prolonged pregnancy. The assay of estriol has the widest application. (150 refs.) - *S. Glinsky.*

University of Aberdeen
Aberdeen, Scotland

2496 **HEIKKILA, JORMA; & LUUKKAINEN, TAPANI.** Urinary excretion of estriol and 15α-hydroxyestriol in complicated pregnancies. *American Journal of Obstetrics and Gynecology*, 110(4): 509-521, 1971.

Estriol and 15α-hydroxyestriol (estetrol) usually give the same information about fetal condition, but in certain conditions only estetrol determinations are reliable. Urinary excretion of estriol and estetrol was determined in 89 women whose pregnancies were complicated by toxemia, diabetes mellitus, Rh isoimmunization, intrauterine fetal death, or recurrent intrahepatic cholestasis of pregnancy. A total of more than 300 urine samples were analyzed by a highly specific gas chromatographic method and the results compared with values during normal pregnancies. Although the 2 estrogens usually furnish the same correct information on the fetal state, estetrol was more reliable in some cases of toxemia. In Rh isoimmunization pregnancies the urinary excretion of estetrol bore a linear correlation to the cord blood hemoglobin concentration, while estriol showed no correlation. (28 refs.) - *S. Glinsky.*

University of Helsinki
Helsinki, Finland

MEDICAL ASPECTS – Prevention and Etiology (General)

2497 Work with reimplanting ova moves another step ahead. *Journal of the American Medical Association*, 215(1): 27-28, 1971.

Recent breakthroughs in successful culturing of human ova fertilized *in vitro* have broad implications for study of the very early stages of human development. Further experiments may lead to the isolation and control of those factors which cause such congenital anomalies as mongolism, dysfunction, and various chromosome abnormalities. So far 5 eggs, aspirated from the ovaries of 3 women, have been fertilized and successfully cultured to the blastocyst stage. Karyotyping at this point has revealed all 5 to be normal diploid cell formations, but reimplantation in the uterus has yet to be attempted. (No refs.) - *N. Mize.*

2498 **RUTT, C. N.; & OFFORD, D. R.** Prenatal and perinatal complications in childhood schizophrenics and their siblings. *Journal of Nervous and Mental Disease*, 152(5): 324-331, 1971.

In an effort to analyze the role played by pregnancy and birth complications in the etiology of childhood schizophrenia, 33 hospitalized autistic children (CA 5-4 to 13-10 yrs) and their 83 siblings were studied. Previous investigations of this relationship have yielded conflicting findings, partially attributable to the lack of comparable case populations and to the small sample size, and partially resulting from inconsistencies in data sources. The current study used data from hospital admissions records, hospital birth records, and records filed with the Pennsylvania State Bureau of Vital Statistics, and compared the 2 groups on a number of prenatal and perinatal complications parameters. Analysis of results showed a higher rate of prenatal complications among the childhood schizophrenics, but the difference was not significant. Perinatal complications (breech presentation, placenta previa, respiratory distress, prolonged labor, etc.), however, were significantly more common ($p<.01$) in the affected group. These findings are in line with previous studies which similarly used siblings as controls. The apparent lack of correlation between these same complications and the IQ of the schizophrenic child is still subject to further reevaluation following work with a larger sample size. A clear association was shown, however, between the autistic child's ego status and his IQ and age at admission to hospital, with the child poorer in ego development tending to be admitted at an earlier age and exhibiting a lower IQ ($p<.01$). While it is unlikely that brain damage resulting from reproductive complications can alone explain the etiology of childhood schizophrenia, it is suggested that adverse perinatal factors, particularly, can profoundly disrupt the child's later development in families already genetically predisposed to this condition. (22 refs.) - *N. Mize.*

Hershey Medical Center of the
Pennsylvania State University
Hershey, Pennsylvania 17033

2499 Mass screen for Tay-Sachs carriers. *Medical World News*, 12(19):35, 1971.

A pilot screening program aimed at identifying all at-risk carriers of Tay-Sachs disease has recently been inaugurated in the Baltimore-Washington area. The Tay-Sachs Disease Program, sponsored by the Kennedy Institute and Johns Hopkins University, uses a simple inexpensive blood test to identify carriers and amniocentesis to detect an affected fetus *in utero*. In the latter case, therapeutic abortion is offered. Because this recessive genetic disorder, characterized by an inability to break down gangliosides in the brain due to lack of the hexosaminidase enzyme, affects primarily Ashkenazi Jews, it was judged suitable for such an experimental mass screening program. Community organizations and synagogues publicize the program in the Jewish community, particularly encouraging Jewish couples of childbearing age to be tested. Costs of the program are estimated at $100,000 to $200,000,

a sum less than that incurred in caring for just 2 Tay-Sachs victims during their short lives. (No refs.) - *N. Mize.*

2500 **WEHRLE, PAUL F., ed.,** Report of the Committee on Environmental Hazards and Subcommittee on Accidental Poisoning of Committee on Accident Prevention, American Academy of Pediatrics. Acute and chronic childhood lead poisoning. *Pediatrics*, 47(5): 950-951, 1971.

In order to promote effective action in the eradication of acute and chronic childhood lead poisoning, the American Academy of Pediatrics recommends that major emphasis be placed on preventive action (testing of dwellings for lead-pigment paint in order to identify high-risk areas) and on determining blood lead values of all 12- to 15-month-old children living in these high-risk areas. Children with blood lead values greater than 50 μg/100 ml should be referred immediately for medical evaluation and repeat determinations. When 2 samples from an individual show concentrations greater than 50 μg/100 ml the responsible local governmental agency should receive reports so that it may investigate and take appropriate action. The true incidence of plumbism is not known; however, 10 to 25% of the children living in deteriorated urban slum housing show evidence of increased lead absorption, and 2 to 5% show evidence of poisoning. Approximately 1/3 of the survivors of acute lead encephalopathy have permanent, irreversible brain damage. (No refs.) - *M. S. Fish.*

No address

2501 **EDWARDS, ROBERT G.; & SHARPE, DAVID J.** Social values and research in human embryology. *Nature*, 231(5298): 87-91, 1971.

Legislative restrictions and social attitudes have, until recently, greatly restricted research in human embryology. In spite of recent changes in both the laws and attitudes, intervention into fetal development is still restricted to artificial insemination, abortion of unwanted fetuses, and transfusing rhesus fetuses. Now the rapidly growing ability to fertilize human eggs in culture, grow them in the laboratory, and then replace them in the mother requires that the emerging issues be considered in order to define social attitudes on which to base rules of conduct of society and of scientists. These new studies may result in the alleviation of certain types of infertility, in determining the sex of the embryo, and in modifying embryos. Present laws are unprepared to deal with the complex issues arising from possible results of this type of experimentation, and perhaps the imposition of additional public statutes is not the best present approach. Instead of authority, control, and bureaucracy, a better solution of the problem might be the establishment of an organization to which biologists and others could come for advice and guidance. Such an organization should be composed of experts whose achievements and understanding would provide a reliable source of opinion and information. (28 refs.) - *M. S. Fish.*

University of Cambridge
Cambridge, England

2502 Rules and regulations for test-tube babies. *Nature*, 231(5298): 69-70, 1971.

A considerable number of recent experiments with laboratory animals on the study of the reproductive process bring up the question as to the propriety of such research and the possible need to guide it. The physiology of reproduction is an important field for the study of contraception and for understanding the nature of genetic defects and of intrauterine life. There are also economic benefits to be gained by learning more about reproduction in domestic animals. But the benefits of genetic engineering in humans are much less obvious. Concern about overpopulation tends to reduce the social impact of infertility in the individual, and philosophical doubts about the goals of human genetics have arisen. Reproductive errors resulting from these experiments, while acceptable to some degree in animals, would constitute major problems in man, particularly if a technique such as cloning were to reproduce such errors manyfold. Needs in this area include a full exchange of information among reproductive physiologists. A professional forum commanding the respect of the scientists working in this field should be available to provide advice; the exchange of information might well be monitored by people outside the field of reproductive physiology. (No refs.) - *M. S. Fish.*

2503 Exploring the genetics of mice and men. *Medical World News*, 11(20): 26-27, 1971.

Cell hybrids have been discovered as being remarkable tools for studying human genetics. Experiments have been done which yield clues to the mechanisms regulating gene expression, and to mapping of genes in order to find which chromosomes genetically transmit diseases such as the Lesch-Nyhan syndrome and a number of blood disorders. It may become possible to correct genetic defects in somatic cells by inserting small amounts of foreign genetic material into them. Cells from an individual could be grown, corrected without introducing foreign antigens (which cause rejection), and then administered to the patient. The homograft reaction will first have to be eliminated before this is possible. (No refs.) - *T. C. Turner.*

2504 To screen or not to screen? *Journal of the American Medical Association*, 215(12): 1978-1979, 1971.

Routine batteries of laboratory screening tests have been criticized on the basis of high cost and lack of clinical importance. But the finding of an abnormal value has great importance in the prevention of complications even if the underlying process cannot be reversed. The introduction of automation has made the cost of screening reasonable, and the discovery of a previously unsuspected abnormality must surely be worth it to the individual involved. (3 refs.) - *E. L. Rowan.*

2505 **MITCHELL, ROSS G.** The prevention of cerebral palsy. *Developmental Medicine and Child Neurology*, 13(9): 137-146, 1971.

Although the present emphasis in cerebral palsy research is on therapy and social support, this disorder is amenable to primary, secondary, and tertiary prevention. Primary measures in the protection of the growing brain include good antenatal and obstetric care. The prevention of hypoxia, hypoglycemia, hyperbilirubinemia, and hypernatremia in the neonatal period and of infections and traumatic insults after the neonatal period are secondary measures. Tertiary prevention of superimposed disabilities includes physical therapy, special education, vocational training, and general management. While the cost of such preventive measures on a national scale will be great, the benefits will be greater. (20 refs.) - *E. L. Rowan.*

University of Aberdeen
Foresterhill, Aberdeen AB9 2ZD, Scotland

2506 **ROGERS, MICHAEL G. H.** The early recognition of handicapping disorders in childhood. *Developmental Medicine and Child Neurology*, 13(1): 88-101, 1971.

A review of the theoretical and practical aspects of early recognition of handicapping disorders in children in the United Kingdom is presented. An examination of the rationale of early identification indicates that: early identification is a sound basis for the management of childhood handicaps; search for delay in development of children should be part of the larger orientation to community child health; more training in developmental medicine is necessary to utilize the techniques for developmental assessment which now exist; surveillance is preferable to screening in developmental procedures; and the risk register concept is unsound and should be replaced by the concept of "child at risk" for clinical use. Early identification should include whole population surveillance carried out as a community health service by competent physicians and health visitors. The child health clinics are the focal points, and assessment centers for referring children are an essential backing to these clinics. The hospital pediatrician should be responsible for selection of the "high risk" group, and coordination of effort should insure that delays in referral are reduced, that relevant and accurate data on a child's development are available for clinical use, and that efforts are made to avoid giving parents conflicting opinions and advice. The lack of adequate numbers of trained personnel and the need for improved services for handicapped children both complicate the manpower problem. (74 refs.) - *M. S. Fish.*

Reading County Borough Health Department
Reading RG1 1JB, England

2507 **INDYK, LEONARD; & COHEN, SHEP.** Newborn intensive care in the United States, east and west. *Clinical Pediatrics*, 10(6): 320-327, 1971.

Following a survey of 12 Newborn Intensive Care Units (NICU), a systematized method and a scoring system (analogous to the APGAR system for newborns) were developed for estimating quality of care in these intensive-care units for seriously ill infants, and 15 separate categories for evaluating NICUs were defined. Most of the units were directed by a full-time pediatrician, often assisted by an anesthesiologist. Nursing coverage ideally had at least one nurse for 2 critically ill infants. Design of units varied from a circular set of rooms surrounding a central nursing station to a linear arrangement with a nursing station at one end. Oxygen was provided at each incubator (portable radiant heaters are useful in the incubator). An X-ray machine was usually brought from a central radiology department. Sophisticated monitoring devices (for body temperature, blood pressure, heart rate, respiration rate) are essential, particularly in detecting apnea (an electric-shock apparatus has been developed for stimulating apneic infants). A variety of devices was available for ventilatory support. Statistical analysis of mortality and morbidity data is needed for complete evaluation of NICUs. (8 refs.) - *B. Berman.*

Children's Hospital
Denver, Colorado

2508 SWITKES, DANIEL A. Cost benefits of neonate screening. *New England Journal of Medicine,* 285(4): 239-240, 1971. (Letter)

The screening of 374,341 newborn infants for galactosemia was reported in "Galactosemia Screening of Newborns in Massachusetts," by Shih et al. (284(April 8):753-757, 1971). This screening represents an irresponsible waste of money, effort, and personnel. Only 1 case and 1 presumptive case of galactosemia were found. The general health of the population, particularly the poor, is neglected while researchers pursue such esoteric subjects at large medical centers. (No refs.) - *K. L. Woodworth.*

No address

2509 SHIH, V. E.; & LEVY, H. L. Cost benefits of neonate screening. *New England Journal of Medicine,* 285(4): 240, 1971. (Letter)

The Massachusetts Metabolic Disorders Screening Program, of which galactosemia screening is a part, detects disorders causing MR and other illnesses. Tests on 2 specimens of blood and a urine specimen from each infant are performed, with galactosemia testing performed on the same blood specimen collected for other tests. It therefore creates no additional burden for the staff. Costs are $1.75 per infant for the entire program, with $0.20 of this spent on the galactosemia testing. The low incidence of galactosemia found could still be due to chance, since it is higher in other states and in several European countries. However, the cost of the screening program has been repaid by the savings to society in terms of the prevention of institutionalization of persons who would have been MR due to such diseases as PKU. The cost of a program screening for galactosemia only would be considerably more than $0.20 per infant, but the establishment of a program testing for only one rare disease is not recommended, for it would be wasteful and inefficient. (No refs.) - *K. L. Woodworth.*

State Laboratory Institute
Boston, Massachusetts

2510 GORDON, JEOFFRY B. Cost benefits of neonate screening. *New England Journal of Medicine,* 285(4): 240, 1971. (Letter)

In reference to the screening of newborns for galactosemia reported by Shih et al., the broader ramifications of such research should be reported. One should be able to assess both the benefits and the costs of such a screening program. Costs are important when resources are limited. Assuming that the marginal costs of the galactosemia screen were minimal, since blood specimens were being collected for phenylketonuria (PKU) testing anyway, it would be appropriate to report total costs of a new program (without PKU testing), the marginal cost of this program, and the cost per case identified. (No refs.) - *K. L. Woodworth.*

University of California
San Diego, California

2511 YOUNGBLUT, ANN. Specially for the newborn – intensive care in the nursery. *Canadian Nurse,* 67(8): 24-27, 1971.

In 1967 Grace General Hospital, Winnipeg, Manitoba, instituted a central neonatal intensive care unit dedicated not only to reducing infant mortality but to helping the infants survive intact by quality care. In keeping with the concept of "family centered maternity care," parents are encouraged to come to the nursery. Babies are taken out of isolettes as soon as their conditions allow, being moved to cribs when their weight reaches 2,040 Gm. At discharge, the public health department is notified, so that there will be some continuity of care. Facilities and equipment consist of 8 wall mounted oxygen and suction outlets, 8 isolettes, a Harco Cardiac Monitor, a Sage infusion pump, phototherapy units, a portable X-ray machine, and a laboratory. Nursing staff receives ongoing inservice education and a qualified pediatrician is in charge, with 7 rotating staff doctors. Conferences are held to adopt new procedures or correct present ones. Any infant who requires special care is admitted. The future goal of the hospital is to admit any infant up to 6 weeks of age with a surgical or medical problem, since measures which prevent death can also prevent brain damage. (6 refs.) - *T. C. Turner.*

Grace General Hospital
Winnipeg, Manitoba, Canada

2512 **SCHMICKEL, ROY; BLOOM, ARTHUR; WORK, BRUCE; & JAFFE, ROBERT.** Amniocentesis in early pregnancy: Prenatal detection of genetic disease. *Michigan Medicine*, 70(16): 611-613, 1971.

Genetic considerations indicate the use of amniocentesis early in pregnancy. Analysis of fetal cells obtained by amniocentesis makes possible the reliable and safe detection of both chromosomal disorders and inborn metabolic errors. In the state of Michigan the price of each diagnosis of mongoloidism in women over 40 would be equal to about 10 months of institutional care for a mongoloid. (5 refs.) - *S. Glinsky.*

University of Michigan Medical School
Ann Arbor, Michigan

2513 **LENZ, W.** Erbberatung bei Schwachsinn (Genetic advice in mental deficiency). *Monatsschrift fur Kinderheilkunde*, 119(5): 191-192, 1971.

The specificity of advice given in genetic counseling depends upon the type of MR involved. Definite risk figures can be supplied in some cases of MR which are due to single genes, such as phenylketonuria or epiloia. Low grade MR is largely determined by polygenes. In high grade MR family planning is desirable for eugenic reasons and for the benefit of the individual family members. (4 refs.) - *S. Glinsky.*

Institut fur Humangenetik
4400 Munster, Federal Republic of Germany

2514 **RAO, B. S. SRIDHARA RAMA; NARAYANAN, H. S.; & REDDY, G. N. NARAYANA.** A clinical and biochemical survey of 729 cases of mental subnormality. *British Journal of Psychiatry*, 118(546): 505-507, 1971.

A clinical and biochemical screening of 729 cases of MR at the Mental Hospital in Bangalore, India, showed a high degree of consanguinity. Comparative observations of MR in 3 rural villages revealed an MR incidence rate of 3.4 per 1,000 and detected 7 cases of phenylketonuria and 2 cases of Hartnup's disease. In all these metabolic defect cases there was a history of consanguinity not only among parents but also among grandparents. (3 refs.) - *S. Glinsky.*

All-India Institute of Mental Health
Bangalore 27, India

2515 **HOOK, E. B.** Monitoring human birth defects and mutations to detect environmental effects. *Science*, 172(3990): 1363-1366, 1971.

A summary is given of the matters discussed at the Birth Defects Institute of New York State Health Department, 1970. The conference was primarily concerend with methods for detecting increases in background rates of birth defects or mutations. Strategies suggested for increasing efficiency of monitoring birth defects were: directed rather than undirected searches; attention to bizarre, gross, and immediate effects of possible environmental insults; immediate attention to the cases observed and the population from which they came; free communication among clinician, scientist, and epidemiologist. Study of fetal wastage is useful, since the rate of malformation

is much higher in abortuses than in newborns, but a difficulty arises in that the total number of conceptuses from which the sample is taken is uncertain. In discussing the interpretation of major common malformations in liveborns as indications of environmental teratogens, anencephaly was suggested as having the best evidence of sensitivity to environmental factors. The use of fresh dominant cases of multiple congenital malformation syndromes as sentinel phenotypes was suggested as a way of monitoring mutation rates. Death certificates might be useful in studying birth defect rates, as well as computerized hospital records and zone electrophoresis of cord blood. Chromosome breakage may also be a sign of a mutagenic insult. A histochemical method using 2-deoxyglucose-6-phosphate as a substrate for detecting variants of glucose-6-phosphate dehydrogenase in leukocytes was discussed as a possible way to monitor somatic cell mutations. A question was raised concerning the problem of confidentiality in record gathering, but the reply was made that the confidentiality of collected information is mandated by law. (No refs.) - *T. C. Turner.*

New York State Dept. of Health
Albany, New York 12208

2516 **LAPHAM, LOWELL W.; & MARKESBERY, WILLIAM R.** Human fetal cerebellar cortex: Organization and maturation of cells in vitro. *Science*, 173(3999): 829-832, 1971.

Cell maturation and the development of a highly organized pattern of growth in organotypic cultures of human fetal cerebellar cortex are described. Explants of cerebellar cortex with leptomeninges intact were prepared from specimens which had been removed surgically for the purpose of terminating pregnancy. The tissue was sectioned and suspended in 15ml of nutrient mixture F-12, then distributed among 5-10 Petri dishes. After a few days cells migrated from the explant in growing numbers. Cells migrating earliest had pale and moderately large nuclei, with broad, sheetlike cytoplasm. These were interpreted as immature astrocytes. Slender neuritic processes had emerged by the end of the first week, followed later by migration of cell bodies from which they arose. Their nuclear size, chromatin pattern, and scanty cytoplasm were characteristic of immature granule neurons. An unexplained finding was the development of unique areas of organization, usually within 3-6 weeks after explantation. The explant was surrounded by a less cellular outgrowth zone which was enclosed by a circular rim of cells (fibroblasts), the rim behaving like a leptomeningeal covering. Large cells suggestive of Purkinje neurons were observed after 4-6 weeks. The primary explants surviving longest were from 18-week-old fetuses. No synapses or myelin were seen in them, and granule cells gradually disappeared from the tips of the neuronal processes, but were a prominent component with the explant proper. Observations suggest that further neuronal maturation in terms of cellular interaction in explants of human fetal cerebellar cortex is most likely to take place within or near the explant proper. The presence of leptomeninges did not seem to interfere with the maturation and cytodifferentiation taking place in the explant and outgrowth zones. The characteristics suggest that regulatory mechanisms which influence developmental phenomena within cells operate *in vitro*. It may be possible, with long-term organotypic cultures, to investigate the processes controlling growth and differentiation in the human fetus. (15 refs.) - *T. C. Turner.*

University of Rochester Medical Center
Rochester, New York 14642

2517 **KASS, LEON R.** What price the perfect baby? *Science*, 173(3992): 103, 1971. (Letter)

Kass comments upon an article in *Science* (January 8, 1971) written by Bentley Glass, in which he discusses the future state of affairs in which babies would be conceived in laboratories and parceled out to foster parents. Glass also said that the paramount right is not to procreate, but the "inalienable right" of every child to be born with a sound mental and physical constitution, based on a sound genotype. Kass asks: whose duty will it be to see that only unblemished children are born and how will this duty be fulfilled? To achieve quality control over human life, human conception and gestation would have to be brought into the laboratory, inspected, tested, graded, and approved. Human reproduction would then be nothing more than manufacture. The question is: Do we want to pay this price? (No refs.) - *T. C. Turner.*

5511A Oak Place
Bethesda, Maryland 20034

2518 **GORDON, HYMIE.** Genetic counseling. Considerations for talking to parents and prospective parents. *Journal of the American Medical Association,* 217(9): 1215-1225, 1971.

Genetic counseling is usually sought either when a child is born with an inherited disease or congenital anomaly and the parents want to know if the condition is likely to recur in their subsequent children, or when a young couple is considering marriage and 1 partner has a relative with a possible heritable disease. The decrease in pediatric disease associated with malnutrition and infection and the increase in the understanding of genetic principles have made genetic counseling a relatively more important part of medical practice. Patterns of inheritance – autosomal dominant, autosomal, recessive, and X-linked – emerge upon inspection of the pedigree and permit estimation of genetic risk. For autosomal dominant, the risk of inheritance is 1 in 2; for autosomal recessive it is 1 in 4; for X-linked recessive, it is 1 in 2 for subsequent sons or 1 in 4 for all subsequent children. Sporadic occurrence of diseases may be due to a mutagenic agent in one parental gonad resulting in development of a fresh mutant gene. Parental consanguinity carries a certain risk, but at the individual level the risk is small. Congenital anomalies are not governed by mendelian genetics, but by polygenic and environmental factors; they can be predicted only in terms of empirical risks. The 3 prerequisites for sound genetic counseling are a precise diagnosis, an adequate family history, and a familiarity with modern genetic principles and developments. Whenever possible, the genetic counselor should enlist the assistance of the family doctor in planning the correct approach to counseling. (15 refs.) - *B. J. Grylack.*

Mayo Clinic
Rochester, Minnesota 55901

2519 **MUKHERJEE, ASIT B.; BLATTNER, PEGGY Y.; & NITOWSKY, HAROLD M.** Sex-chromatin fluorescence in human amniotic-fluid cells. *Lancet,* 2(7726): 709-710, 1971. (Letter)

Although no recognizable fluorescence of the Barr body has been observed in freshly obtained uncultured amniotic-fluid cells from female fetuses collected by amniocentesis, the Barr body and Y-body in different samples of amniotic-fluid fibroblasts that are grown directly on microscope slides do show a distinct fluorescence when stained with quinacrine mustard, and sex identification can be made without ambiguity. Seven of 11 specimens were clearly identified as females and 4 as males by their characteristic fluorescent bodies, and sex-chromosome constitution obtained by chromosome analysis of all the fibroblast samples confirmed the results of the fluorescent study. The incidence of cell nuclei with fluorescent Barr bodies varied from 44% to 74% in different samples, depending upon and increasing with cell density, while that of the fluorescent Y-body varied from 75% to 84%. (7 refs.) - *B. J. Grylack.*

Albert Einstein College of Medicine
Bronx, New York 10461

2520 **McLAREN, HUGH CAMERON.** Abortion in U.S.A. *Lancet,* 2(7726): 713, 1971. (Letter)

Planned Parenthood/World Population (U.S.A.) has established clinics for abortions. Moreover, a physician associated with International Planned Parenthood Federation discusses abortion in his study of contraceptive practice, although he acknowledges that abortion on purely social grounds is illegal. The various family planning agencies in England should not be allowed to follow the American example. (No refs.) - *B. J. Grylack.*

Birmingham Maternity Hospital
Birmingham B15 2TG, England

2521 **VLASOVA, T. A.** New advances in Soviet defectology. *Soviet Education,* 14(1,2,3): 20-39, 1971-1972.

Although significant progress has been made in the Soviet Union in the past 5 years in overcoming certain etiological factors leading to impaired physical and mental development in children, many other factors still exist. Prominent among them are natural trauma, asphyxia, disruptions in the nutrition of the fetus, Rh conflicts, chromosome pathology, certain types of infectious diseases contracted by the mother during pregnancy, and diseases of the child himself, especially during early life. The Scientific Research Institute of the Study of Defects of the U.S.S.R.

Academy of Pedagogical Sciences is the principal institution working on these problems in the Soviet Union. One of the major tasks of the Institute in recent years has been the establishment of a scientific basis for the development of special schools for the separate instruction of handicapped children. The Institute has considerably expanded and intensified its research on the clinical psychological, neurophysiological, and pedagogical study of children with anomalies and on the analysis of structures of defects involving various developmental deficits. The surmounting and prevention of defects in the development of handicapped children has been stressed through the establishment of special programs for handicapped preschool children. (No refs.) - *B. J. Grylack.*

U.S.S.R. Academy of Pedagogical Sciences
Moscow, U.S.S.R.

2522 PEVZNER, M. S. Basic directions in the study of oligophrenia. *Soviet Education,* 14(1,2,3,): 40-48, 1971-1972.

Recent studies have related the problem of oligophrenia to a whole series of areas, including genetics, biochemistry, immunology, and psychology. Soviet research on the personality of the MR individual and his social adaptation has been extensive, and experiments have revealed the diversity of individual mental functions in various stages of learning in children afflicted with oligophrenia. Joint clinicophysiological and psychopedagogical investigations of the structure of MR, together with studies of localized physical defects, have provided a basis for assuming that the diffuse underdevelopment of or damage to the cerebral cortex is the basic pathogenetic factor in oligophrenia, and this hypothesis has been confirmed by experimental studies. It appears that the qualitative uniqueness of the structure of the defect is determined by etiopathogenetic factors and by the time of damage, its location, and its etiology. On this basis, 5 forms of oligophrenia have been delineated: the uncomplicated form, oligophrenia with the cerebrasthenic syndrome, with local injuries, with psychopathological forms of behavior, and with frontal insufficiency. Each of these forms is characterized by specific features relating to the structure of the defect. (1 ref.) - *B. J. Grylack.*

U.S.S.R. Academy of Pedagogical Sciences
Moscow, U.S.S.R.

2523 CAMPBELL, M. A.; & ELSTON, R. C. Relatives of probands: models for preliminary genetic analysis. *Annals of Human Genetics,* 35(Part 2): 225-236, 1971.

In the genetic study of a particular trait, it has been common practice to summarize the data by stating the frequencies with which the trait occurs among the various types of relatives of probands. A comparison of the expected probabilities of being affected along with the reported empirical proportion can provide a clue as to which genetic hypothesis is the most appropriate. If the genetic model is known, the prediction of risk figures for use in counseling can be achieved in a reasonable way. Unknown parameters involved in genetic analyses can be estimated if sufficient data are available. As a clinical example, extensive data on pyloric stenosis have been used in multiple locus models, particularly those involving an X-linked gene. The distorted sex-ratio of 4 affected males to 1 affected female cannot be accounted for by a single major gene hypothesis. Preliminary indications suggest that a multi-locus hypothesis would correspond much better with the available data. (14 refs.) - *B. J. Grylack.*

University of California at Los Angeles
Los Angeles, California 90024

2524 HOAG, ROGER W. Use of urinary estriol determinations in high-risk pregnancy in the community hospital. *American Journal of Obstetrics and Gynecology,* 110(2): 203-209, 1971.

Determination of urinary estriol levels can be useful in the management of high-risk pregnancies in a community hospital, although it is subject to faulty use or misinterpretation. In 199 high-risk pregnancies 24 hour urinary estriol assays were done. The correlation between estriol level obtained within 7 days of delivery and fetal survival appeared similar to previously reported correlations. If estriol was above 16 mg/24 hours no fetal loss occurred past 33 weeks of gestation, and no infants survived when the estriol levels were below 4.9 mg/24 hours. Estriol determinations were helpful in management in 143 cases. The greatest frequency use of the test was in permitting a pregnancy to progress to term. Intervention in 17 cases was based upon test results. The high cost of estriol determinations

and their dependence on several criteria for validity suggest great care in their use. (9 refs.) - *S. Glinsky.*

Department of Gynecology and Obstetrics
Alta Bates Community Hospital, California

2525 Preventing birth defects. *Nature,* 234(5331): 506, 1971.

L'Institut de la Vie (Paris, France) held a meeting in December 1971 to discuss strategy in preventing birth defects and alleviating their physical, psychological, and social effects. Three major aspects of the problem were identified: (1) Lack of reliable statistics in many countries; (2) recognition of new teratogens; and (3) prevention and cure. McBride (Sydney Lying-in Hospital) stated that in New South Wales, Australia, 5% of all children are seriously defective: 1.5% have major physical defects; 1% have cerebral palsy: and 2.5% are MR. Whitehead (Kampala) pointed to the permanent damage to the central nervous system which can be caused by protein malnutrition, particularly if the fetus or young child is malnourished during one of the critical periods of development. Miller (University of British Columbia) called for accurate and comprehensive recording of birth defects, so that unusual patterns can be identified quickly. Hirschhorn (New York University School of Medicine) felt it was now possible in theory to eradicate mongolism by recognizing its karyotype in fetal cells and aborting the fetuses concerned. Fetal cells removed by amniocentesis can be cultured and assayed for enzymatic deficiencies and it might be possible to induce synthesis of enzymes not already present. A point underemphasized was that the biggest single contribution to the eradication of birth defects would be reduction of malnutrition. (No refs.) - *S. M. Rucker.*

MEDICAL ASPECTS – Etiologic Groupings
Infections, intoxication, and hemolytic disorders

2526 Could blacks be prone to lead pollution? *Medical World News,* 12(21):21, 1971.

Comparison between 68 glucose-6-phosphate dehydrogenase (G6PD) - deficient black children and a group of 59 nondeficient black controls matched for age, sex, socioeconomic status and census tract, indicates that this enzyme deficiency, which affects 13% of American blacks, may result in higher blood lead levels. Mean whole-serum lead levels were 0.0299 mg% in the G6PD - deficient children as compared to a mean of 0.0268 mg% in the controls. Red cells tested alone showed an even greater difference between the 2 groups: 0.0469 mg% as compared to 0.0350 mg%. The children all lived within a 1-mile radius of a known lead emission source in Omaha. *In vitro* work suggests that lack of available carbohydrate in the enzyme-deficient group affects membrane permeability, thus leading to a greater lead accumulation. This represents the first evidence of genetic susceptibility to an air pollutant. (No refs.) - *N. Mize.*

2527 **FINKEL, ASHER J.** Measles, a renewed challenge. *Journal of the American Medical Association,* 216(6): 1018, 1971. (Editorial)

While a widespread measles vaccination program was able to reduce the number of reported rubella cases in the United States from 204,136 in 1966 to 22,331 in 1968, there has been a slow rise since this time, largely attributable to the failure of the vaccination program to reach preschool children in urban poverty and rural areas. Since experience with available vaccines has shown their efficacy to exceed 90%, a concerted effort should be made to expand the program to all sectors of the population. It is estimated that between 1963-68, 3,244 cases of MR, a frequent complication of measles, were averted through measles vaccination. A successful program should further reduce and hopefully eradicate the morbidity and death associated with this disease. (6 refs.) - *N. Mize.*

Department of Environmental, Public,
and Occupational Health
Chicago, Illinois

2528 **VELLA, WALLY.** Rubella vaccination. *Lancet,* 1(7710): 1188, 1971. (Letter)

Present policy of the British Army Medical Department is to test all pregnant women seen in the Service's antenatal clinics for the presence of

rubella antibodies. Any pregnant woman found to be seronegative is offered vaccination immediately following parturition, accompanied by medical instruction that she employ suitable contraceptive practices for a subsequent 2 month period. Rubella vaccination is also offered to any seronegative non-pregnant woman who desires protection and to all 11 to 14-year-old girls. (1 ref.) - *N. Mize.*

Royal Army Medical College
London S.W.1, England

2529 **BOUE, ANDRE; NICOLAS, ANDRE; & MONTAGNON, BERNARD.** Reinfection with rubella in pregnant women. *Lancet,* 1(7712): 1251-1253, 1971.

Clinical experience with rubella reinfection in women exposed to the virus during the first trimester of pregnancy has generally indicated no risk to the developing fetus. Three cases are cited here which support this observation. In all 3 women with pre-existing levels of rubella hemagglutination-inhibition antibody, a significant rise in rubella antibody titer was detected following virus exposure, but there were no clinical reactions. A sucrose density-gradient ultracentrifugation procedure determined the rise in titer to be only in the IgG fraction, a characteristic secondary response in the naturally immune S. No interruption of pregnancy was attempted, and the women subsequently delivered normal infants exhibiting no malformations or symptoms attributable to congenital rubella. This experience emphasizes the importance of distinguishing between instances of primary response during a subclinical infection and the relatively harmless instances of reinfection in cases where a significant rise in rubella antibody titer is detected in the pregnant woman. Such procedures can help reduce the number of therapeutic abortions performed unnecessarily. (5 refs.) - *N. Mize.*

Laboratoire S.E.S.E.P.
Chateau de Longchamp
75 Paris 16, France

2530 Screening for lead: Quick and easy test points for plumbism. *Medical World News,* 12(17): 19, 1971.

A quick and simple blood test for detecting high lead levels in children has so far proven 85-95% effective. The test, based on the fact that lead-bound erythrocytes do not hemolyze as easily as normal red cells in a hypotonic solution, can be conducted in less than 30 minutes and requires only routine lab equipment. Tests conducted on normal adults and children, and on children with proved plumbism, have supported the assumption that the lower the hemolysis the higher the lead level. Advantages of this new indirect screening technique over previous methods include the need for less blood, the shorter time period required to obtain results, and the relatively inexpensive lab equipment involved. While there appears to be some overlapping between lead intoxication results and those for sickle cell disease and thalassemia, the test does provide the quick, accurate indication of lead levels necessary to begin immediate chelation therapy for the estimated 2 - 5% of urban children suffering from serious lead poisoning. (No refs.) - *N. Mize.*

2531 DPH detected in human neonates. *Medical World News,* 12(11): 88E, 1971.

The passage of diphenylhydantoin (DPH) across the placenta has been demonstrated in prospective clinical studies of 7 epileptic mothers. Maternal plasma levels of the drug at term ranged from 1.0 to 6.3 μg/ml despite the relatively uniform dosage level of 300 mg of DPH per day in 6 mothers and 400 mg per day in 1 mother. Clearance of the placentally transferred DPH was virtually complete by the fourth or fifth postpartum day. A plasma half-life for DPH was calculated to be about 60 hours. Although evidence of a teratogenic effect from DPH in the human has not been established, a higher-than-normal incidence of cleft palate and/or cleft lip in the infants suggests such a possibility. (No refs.) - *J. C. Moody.*

2532 **MIZUTANI, HIROMISHI; MIZUTANI, HIROKO; SAITO, S.; NIHEI, K.; & IZUCHI, T.** Cellular hypersensitivity in subacute sclerosing panencephalitis. *Journal of the American Medical Association,* 216(7): 1201-1202, 1971. (Letter)

Although several investigators have confirmed the relation of measles virus to subacute sclerosing

panencephalitis (SSPE), the question of why the disease occurs only rarely among patients with measles has given rise to a divergence of opinion among researchers. A recent study was conducted to evaluate cellular immunity in a case of SSPE. Donors were injected intradermally with 0.1 ml of skin test antigen (a concentrated, purified suspension of the Edmonston strain of measles virus) and macrophage migration inhibition tests were carried out. As a result, the SSPE patient showed a positive migration inhibition test with concentrated lymphocytic supernatant and with concentrated cerebrospinal fluid (CSF). Also, skin tests showed positive. It is posited, therefore, that a positive migration inhibition test with concentrated CSF may have some bearing on the cause of SSPE, and this phenomenon bears investigation. (6 refs.) - *T. C. Turner.*

No address

2533 **JONES, PHILLIP H.** Herd immunity in rubella. *Journal of the American Medical Association,* 216(7): 1202, 1971. (Letter)

It is incorrect to say that no one has seen "herd immunity" in rubella. Adult Americans reach childbearing age without having contracted the disease; this shows that they have been personally protected from infective exposure by herd immunity. The present problem of adult susceptibility in the U.S. is the result of recurrent rubella epidemics not having infected all those susceptible to it. (No refs.) - *T. C. Turner.*

2534 **SCHIFF, DAVID; ARANDA, JACOB V.; CHAN, GEORGE; COLLE, ELEANOR; & STERN, LEO.** Metabolic effects of exchange transfusions. I. Effect of citrated and of heparinized blood on glucose, nonesterified fatty acids, 2-(4-hydroxybenzeneazo)-benzoic acid binding, and insulin. *Journal of Pediatrics,* 78(4): 603-609, 1971.

A study of metabolic effects of exchange transfusions with citrated and heparinized blood has disclosed both advantages and hazards to the use of either in the treatment of neonatal hyperbilirubinemia. The study population consisted of 35 infants on whom 41 exchange transfusions were carried out during the first 6 days of life. Of this group 23 Ss received 26 transfusions with acid citrate dextrose (ACD) blood and 12 received 15 transfusions with heparinized blood. The clinical classification of the 2 groups receiving ACD and heparinized blood, respectively, was: Rhesus affected, 11 and 4; ABO incompatibility, 4 and 1; prematurity, 6 and 6; and other, 2 and 1. Exchanges were performed via the umbilical vessel route to a total volume of 150 ml/kg body weight. Determination of levels of glucose, insulin, nonesterified fatty acids (NEFA), and bilirubin-albumin binding capacity before, during, and after transfusions showed that changes (mean ± S.E.M.) following heparinized and ACD blood, respectively, were: NEFA (meq/1), 1.7 ± 0.3 and -0.5 ± .08; glucose (mg %), -16.2 ± 7.6 and 74.0 ± 4.3; binding (%), 8.2 ± 4.8 and 17.7 ± 1.8; insulin (Mμg/ml), 1.1 ± 2.7 and 22.5 ± 4.8. Although a number of advantages are associated with the use of heparin over ACD as an anticoagulant for blood used in these transfusions, the study has demonstrated 2 potential hazards: low glucose concentration in heparinized blood may not correct, but may potentiate, the hypoglycemic state in some cases of erythroblastosis, and the rise in NEFA with the potential uncoupling effect on bilirubin-albumin binding is disturbing. The use of ACD blood, however, results in an increase in insulin and can cause severe delayed hypoglycemia several hours following transfusion. (24 refs.) - *M. S. Fish.*

Montreal Children's Hospital
Montreal 108, Quebec, Canada

2535 **AULD, PETER A. M.** Oxygen therapy for premature infants. *Journal of Pediatrics,* 78(4): 705-709, 1971.

Management of a case of hyaline membrane disease in a premature infant which required oxygen therapy represents an example of the problems associated with this form of treatment. Concentrations up to 100% oxygen were required for several days after birth. The concentrations were regulated hourly to those which would just abolish cyanosis. The infant was monitored by serial determinations of pH and PCO_2. Improvement occurred by the fifth day, and oxygen concentration was gradually reduced to room air by day 17. Tachypnea persisted, and a chest roentgenogram on day 25 showed markings typical of the Mikity-Wilson syndrome or bronchopulmonary dysplasia. The child was discharged at

6 weeks but was readmitted at 16 weeks with congestive heart failure due to cor pulmonale. The findings were believed to be due to the toxic effects of oxygen. After treatment for congestive heart failure the S improved and at 2 years of age required no more medication. The case typifies the problems of the physician who must treat infants with respiratory insufficiency by providing adequate oxygen while minimizing the possible toxic effects. Present limitations of monitoring the partial pressure of oxygen in blood indicate the need for close clinical observation. (25 refs.) - *M. S. Fish.*

525 East 68th Street
New York, New York 10021

2536 McCRACKEN, GEORGE H., JR. Changing pattern of the antimicrobial susceptibilities of *Escherichia coli* in neonatal infections. *Journal of Pediatrics,* 78(6): 942-947, 1971.

Gentamicin appears to be suitable as initial therapy for suspected or proved systemic neonatal coliform infections. Studies of *in vitro* antimicrobial susceptibilities of 520 *E. coli* from 7 hospitals, utilizing a modified agar plate dilution method and determining the lowest concentration of antibiotic which completely inhibited bacterial growth after incubating for 18 hr. at 37°C, disclosed that the concentration and percent of strains inhibited by 5 antibiotics were, respectively: gentamicin (5 μg/ml), 92-100% of all strains; kanamycin (10 μg/ml), 66-86% of all strains tested; streptomycin (10 μg/ml), 50-81%; ampicillin (10 μg/ml), 65-93%; and polymyxin B (5 μg/ml), 87-100%. The results of the investigation, which was initiated when 5 newborn infants with *E. coli* meningitis failed to respond to kanamycin therapy, emphasize the need for constant surveillance for possible changing antimicrobial susceptibilities. (7 refs.) - *M. S. Fish.*

5323 Harry Hines Blvd.
Dallas, Texas 75235

2537 MURPHREE, ODDIST D.; MORGAN, PAUL N.; & JARMAN, RUTH V. Learning deficits and activity changes: A partial laboratory model in postencephalitic rats for studies of brain damage. *Conditional Reflex,* 6(1): 30-35, 1971.

Hyperactivity and learning disabilities may be related to early brain damage. Rats ranging from 8 to 21 days of age were inoculated in the right cerebral hemisphere with St. Louis encephalitis virus, Japanese B encephalitis virus, and Murray Valley encephalitis virus. Mumps virus was injected at age 1 day. Very young rats usually developed fatal encephalitis, but rats injected at ages of more than 2 weeks showed little or no clinical signs of infection. Inoculation at ages between these extremes resulted in clinical disease in most of the rats, but many survived. Physically, most of these surviving animals appeared to be normal, but when tested at maturity in a complex maze, they exhibited a marked reduction in learning ability. Tilting cage type activity measurements taken overnight with the Murray Valley group also indicated hyperactivity in most of the postinfectious rats. These data imply that such a laboratory model might be related to similar hyperactivity and learning disabilities seen in certain types of minimally brain damaged children. A viral-sex interaction often seen in earlier studies is also noted in our animals injected at younger ages. (8 refs.) - *S. Glinsky.*

Veterans Administration Hospital
Little Rock, Arkansas

2538 KOLER, R. D.; JONES, R. T.; WASI, P.; & POOTRUKUL, S. Genetics of hemoglobin H and α-thalassemia. *Annals of Human Genetics,* 34(4): 371-377, 1971.

Comparison of two models proposed to explain the genetics of the α-thalassemias has shown that both fit observed data from Thailand. The disorders include Hb H disease and Hb Bart's hydrops fetalis. The former results in a partially compensated hemolytic anemia; the latter is lethal *in utero.* The disorders are apparently due to a deficit in human hemoglobin α-chain production, resulting in a relative excess of γ-chains during fetal life and an excess of β-chains in adult life. The one-locus model with 3 alleles presupposes a mild α-thalassemia allele (t) which results in a slight deficiency of alpha-chain production and a second, severe allele (t') causing even greater deficiency. The phenotype/genotype groups, based on increasing percent of Hb Bart's at birth, would be (where T is a normal allele): normal (T/T), silent carrier (T/t), α-thalassemia trait (T/t'; t/t), Hb H disease (t/t'), and hydrops

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fetalis (t'/t'). The two-loci model without linkage makes the same assumptions as the first model but assigns the mild allele to one locus and the severe allele to a second, unlinked locus. Additional studies are expected to distinguish between these two models and a third alternative. (41 refs.) - *M. S. Fish.*

University of Oregon Medical School
Portland, Oregon 97201

2539 **AICKIN, D. R.; & CAMPBELL, D. G.** Anomalous amniotic fluid bilirubin after hepatitis in an Rh immunized patient. *Obstetrics and Gynecology*, 37(5): 687-688, 1971.

Evaluation of a recent case of a jaundiced mother with elevated amniotic fluid bilirubin whose child was not severely Rh immunized indicates that amniotic fluid bilirubin levels should be interpreted with caution when assessing an Rh immunized fetus. The S was a 25-year-old pregnant woman who was referred at 23 weeks of gestation for management of an Rh immunized pregnancy. The S became jaundiced at 28 weeks, and her urine contained bile. Liver function tests showed total and conjugated bilirubin to be 3.1 and 2.4 mg/100 ml, respectively. The patient had recovered from the illness, which was diagnosed as infectious hepatitis, by 32 weeks of gestation; the jaundice had disappeared; and liver function tests gave total and conjugated bilirubin levels of 0.7 and 0.6 mg/100 ml, respectively. Rh antibody titer was 1:32. Amniocentesis revealed markedly yellow fluid; however, spontaneous labor 24 hrs after amniocentesis produced a male infant who, although slightly jaundiced, did not require an exchange transfusion. The findings indicate that elevated bilirubin levels in the amniotic fluid may persist after clinical maternal jaundice has cleared. (4 refs.) - *M. S. Fish.*

Royal Women's Hospital
Parkville 3052, Victoria, Australia

2540 **LEVY, GERHARD; & ERTEL, INTA J.** Effect of bilirubin on drug conjugations in children. *Pediatrics*, 47(5): 811-817, 1971.

Studies of two sisters with phenobarbital responsive hyperbilirubinemia have indicated that their disorder may involve a deficiency in hepatic anion-binding protein, rather than in glucuronyl transferase. The Ss, 2 female siblings of 3 and 9 years of age, were jaundiced since birth; however, they had apparently normal liver function and no abnormal neurological or physical characteristics other than jaundice. The family had no known history of jaundice. Serum direct and indirect bilirubin concentrations and levels of gluconorides of salicylate, salicylamide, and acetaminophen were determined for the Ss and their parents after oral administration of the unconjugated materials. Results indicated that gluconoride formation was normal at widely different to 37 mg/100 ml) serum bilirubin concentrations. The occurrence of normal levels of conjugation of salicylate with glycine and of formation of salicylamide and acetaminophen sulfates also indicated that the biotransformation of these drugs was not affected by the unconjugated hyperbilirubinemia. The clinical and pharmacokinetic evidence is consistent with a deficiency of the hepatic cytoplasmic organic anion-binding protein (also induced by phenobarbital) rather than or in addition to glucoronyl transferase activity. (37 refs.) - *M. S. Fish.*

State University of New York at Buffalo
School of Pharmacy
Buffalo, New York 14214

2541 **THALER, M. MICHAEL; & SCHMID, RUDI.** Drugs and bilirubin. *Pediatrics*, 47(5): 807-810, 1971. (Commentary)

Advances in the area of physiological detoxification processes and of the effect of drugs on hepatic function have provided new approaches to the treatment of hyperbilirubinemia; however, since some of the drugs known to be useful in this treatment are potentially toxic and since their effects on the developing fetus are still largely unknown, they must be employed with caution. Two types of hyperbilirubinemia have been suggested: type I, which is unresponsive to drug treatment; and type II, which is responsive to this approach. The latter type is less severe clinically and likely involves cases of unconjugated bilirubin in which the defect in the enzyme, UDP-glucoronyl transferase, is incomplete and is subject to induction by certain drugs. Phenobarbital, a potent inducer of microsomal enzymes, has been found useful in the treatment of the Crigler-Najjar syndrome, a type of congenital unconjugated hyperbilirubinemia; however,

experimental data indicate that the effect of the drug may be related to other factors as well as to enzyme induction. Other studies have questioned why some types of hyperbilirubinemia respond to phenobarbital, whereas no enhancement is detected in the biotransformation of other compounds. In the use of drugs for this disorder caution must be exercised, since substances such as acetylsalicylic acid are known to impair the ability of albumin to conjugate with bilirubin. (38 refs.) - *M. S. Fish.*

University of California
San Francisco, California 94122

2542 JONSSON, MARIANNE; & ALVIN, ALLAN. A 12-year review of acute bacterial meningitis in Stockholm. *Scandinavian Journal of Infectious Diseases,* 3(2): 141-150, 1971.

A 12-year review of hospital charts of 476 patients with bacterial meningitis has shown that the highest fatality rate was among the newborn with E. coli/coliform and L. monocytogenes infections, and that the frequency of sequelae from all infections was 19.1%, with hydrocephalus and mental defects the most common residues in the children. Charts of the patients, three-fourths under 15 years of age and one-third less than 1 year of age, were examined over 2 periods of time: 1956-1962 and 1963-1967. Two-thirds had Hemophilus influenzae, meningococcal, or pneumococcal infections. The remainder had infections with E. coli or coliform, streptococci, S. aureus, Listeria monocytogenes, myobacterium tuberculosis, or cryptococci. In 18.1% of the patients no pathogen was isolated from the cerebrospinal fluid; in 29.7% of the patients treated with antibiotics prior to admission cultures were negative, and this finding occurred twice as frequently in these patients as in those not previously treated. Blood cultures were positive in 59% and 29% of the fatal cases and the survivors respectively. The fatality rate decreased from 23% in the first period to 13% in the second period. The highest fatality rate observed among the newborn decreased markedly during the second period. (34 refs.) - *M. S. Fish.*

Roslagstulls sjukhus
Box 5901
S-114 89 Stockholm, Sweden

2543 NORDENFELT, ERIK; ANDREN-SANDBERG, MARGARETA; LUNDQUIST, ALF; & HENRIKSON, HANS. Persistence of Australia (Au-) antigen among patients in a hospital for mentally retarded and its relation to chronic liver disease. *Scandinavian Journal of Infectious Disease,* 3(2): 103-107, 1971.

Of a group of hospitalized MR patients, 8% (all male) were found to have the Australia (Au-) antigen in their sera. The Ss were 442 male and 155 female patients hospitalized for severe oligophrenia. These and 688 hopsital personnel were examined for Au-antigen and antibodies against Au-antigen, utilizing the double immunodiffusion and the counter-electrophoresis techniques. Of the group, 36 male patients were Au-positive. No female patients or staff were Au-positive, although 1 each had antibodies. Examination of liver biopsy specimens from 22 Au-positive and 12 Au-negative patients and necropsy specimens from 1 Au-positive and 9 Au-negative patients showed that none of the livers of the Au-positive patients were normal. The diagnosis was chronic persistent and chronic aggressive hepatitis for 15 and 6 patients, respectively. One each had cirrhosis and steatosis. Livers of the 21 Au-negative patients were normal in 16 and had mild or moderate signs of steatosis in 5. (24 refs.) - *M. S. Fish.*

Inst. Med. Mikrobiologi
Solvegatan 23
S-223 62 Lund, Sweden

2544 AHO, KIMMO; JARENFELT, MARKKU; & RAPOLA, JUHANI. Toxoplasmosis as a cause of foetal death. *Scandinavian Journal of Infectious Diseases,* 3(1): 55-57, 1971.

Results of a study of 523 mothers with stillborn infants showed that only 4 mothers had blood specimens with a toxoplasma dye test of 256 or higher, and in only 1 case did autopsy findings in the infant suggest a toxoplasma infection. Examination of blood specimens taken from the 532 mothers immediately after delivery, utilizing the microtiter technique to measure complement-fixing (CF) antibodies, followed by a dye test for the positive sera, showed that 1 of the samples had a CF and dye test titer of 8 and 256, respectively; 2 had comparable values of 32 and

1024, respectively; and 1 had values of ⩾ 64 and 4096, respectively. Only in the latter case were the histological findings consistent with intrauterine toxoplasmosis. In this case the slightly macerated fetus weighed 1,500 g and had a soft and friable brain with small foci of calcification. No toxoplasma organisms were found in free or cystic form. The results indicate that a toxoplasma infection accounted for less than 0.5% of this group of fetal deaths. (9 refs.) - *M. S. Fish.*

State Serum Institute
Helsinki, Finland

2545 VESIKARI, TIMO. Rubella antibodies in infants whose mothers had rubella during the second and third trimesters of pregnancy. *Scandinavian Journal of Infectious Diseases,* 3(1): 1-6, 1971.

Of a group of 16 infants whose mothers had rubella during the second and third trimesters of pregnancy, only 1 infant had permanent rubella antibodies and was chronically ill since birth; all others were clinically normal. Examination for rubella antibodies of serum samples collected from 16 babies and from their mothers showed that in 11 babies the antibodies disappeared within 6 months after birth, indicating that the infants likely escaped intrauterine infection. Antibody titers declined to low levels within 1 year in 2 babies, but in 3 the antibodies remained unchanged or increased slightly during the first year of life, indicating that the infants appeared to have intrauterine virus infections at a later stage of pregnancy. In some infants total IgM antibodies were slightly increased, but no rubella-specific IgM antibodies were found in any of the infants. No rubella virus could be isolated from any of the babies. The one infant who was chronically ill from birth, with a disorder resembling chronic encephalitis, likely had a chronic rubella infection. (16 refs.) - *M. S. Fish.*

University of Helsinki
Helsinki 29, Finland

2546 STONE, DAVID; MATALKA, EDWARD; & PULASKI, BARBARA. Do artificial sweeteners ingested in pregnancy affect the offspring? *Nature,* 231(5297): 53, 1971.

The observation of abnormal behavior in rats treated with cyclamate and in their offspring and a study of the effects of artificial sweeteners on human offspring suggest but do not demonstrate causal relationships in the human. Behavior of rats treated with 20 mg/kg/day of cyclamate was abnormal; their offspring were hyperactive and slow to develop responses to food rewards. Questionnaires returned by 975 women who had given birth to normal children and by women who had given birth to a total of 247 MR children indicated that, in comparing children of the same age group, a statistically significant difference existed in the ingestion of artificial sweeteners by women of the two groups. Among the normal women usage of the sweeteners increased from 11.5% to 14.5% to 21.5% for the periods of 1959-61, 1962-64, and 1965-69, respectively. In the MR group the corresponding figures were 17.7%, 22.6%, and 35.8%, respectively. The data indicated that the ingestion of artificial sweeteners was equally related to both Down's syndrome and other forms of MR. A possible relationship also appeared between ingestion of the sweeteners before and during pregnancy and the incidence of abnormalities in the non-MR group. Overall incidence of behavioral problems was 5.4% and 2.0% for the user and nonuser groups, respectively. The most common problems were hyperactivity, nervous irritability, and extreme nervousness. Physical abnormalities, mainly bone deformities, were reported for 9.7% and 4.4% of the children in the user and nonuser groups, respectively. (2 refs.) - *M. S. Fish.*

Worcester Foundation
for Experimental Biology
Shrewsbury, Massachusetts

2547 Rubella reinfections. *Lancet,* 1(7712): 1279-1280, 1971.

Virological studies have shown that reinfection by rubella virus may occur and that the frequency of this event is greater among cases of vaccine-induced immunity than among those who had a natural infection. One study demonstrated serological evidence of reinfection in 23% of a group of 22 children who had been vaccinated, compared to only 1 comparable case from the group of 66 children who had acquired the immunity naturally. In a military camp only 5 of 149 naturally immune Ss were reinfected, compared to 12 of 15 who had been vaccinated.

Virus was detected in only one of this latter group. Reinfection appears to be related to low HI antibody titers, and these are approximately 4 to 8 times lower in vaccinated compared to naturally immunized Ss. The principal problem centers on the possible hazards in the reinfection of women during pregnancy; however, viral transmission to the fetus likely must occur before the fetus is at risk, and in 3 cases of reinfection of pregnant women during early pregnancy, the infants were clinically normal initially with no evidence of congenitally acquired infection. In cases of maternal subclinical infection serum fractionation studies should be performed in order to determine whether an immune response is primary or secondary. (12 refs.) - *M. S. Fish.*

2548 **RANE, ANDERS; LUNDE, PER KNUT M.; JALLING, BIRGITTA; YAFFE, SUMNER J.; & SJOQVIST, FOLKE.** Plasma protein binding of diphenylhydantoin in normal and hyperbilirubinemic infants. *Journal of Pediatrics*, 78(5): 877-882, 1971.

A comparison of plasma protein binding of diphenylhydantoin (DPH) in normal and hyperbilirubinemic infants has shown that in the affected infants a correlation exists between unbound DPH and total bilirubin concentration, suggesting that DPH and bilirubin compete for binding sites on the albumin molecule. Samples of plasma prepared from fresh cord blood from 13 full-term, normal infants and from 20 hyperbilirubinemic infants and fresh adult plasma from a blood bank were measured for bilirubin, total plasma protein, and plasma protein fractions, including albumin. Samples were also incubated for 30 minutes with labeled (^{14}C) DPH (16 μg/ml final concentration) and assayed for radioactivity. Results showed that the unbound fraction of DPH from the plasma of normal infants was 10.6 ± 1.4%, a value considerably higher than that obtained from adult plasma (7.4 ± 0.7%). Binding capacity of infant cord blood was comparable to that obtained from 2 normal 1- and 18-day-old infants. In the samples from 20 hyperbilirubinemic children, which had total bilirubin concentrations of 4.5 to 24.5 mg/100 ml, a correlation between % or unbound DPH and total bilirubin concentrations was observed ($r = 0.55$; $p<0.05$), and at bilirubin concentrations of 20 mg/100 ml, unbound DPH was twice as high as in plasma from unaffected infants. The correlation coefficient became even greater ($r = 0.69$; $p<0.001$) when the amount of unbound DPH was related to the quotient between total plasma bilirubin and albumin concentrations. (19 refs.) - *M. S. Fish.*

Karolinska Institutet
Stockholm, Sweden

2549 Rubella shots: Warn the women. *Medical World News*, 12(24): 17, 1971.

The dangers of conception too soon after rubella vaccination in susceptible women are underscored by recent evidence that the rubella vaccine virus may well cross the placental barrier. The case is reported of a 25-year-old woman who conceived 6 weeks after vaccination and was aborted 2 months later because of rising antibody titers. Characterization studies of rubella virus, isolated from the eye of the fetus, are expected to confirm that the virus is of vaccine origin and not the result of an undetected natural infection. Greater care needs to be exercised by physicians in the assessment and handling of female vaccinees of childbearing age. Though still in the experimental stages, one program would inject women found susceptible to rubella with a long-lasting contraceptive, such as medroxyprogesterone acetate, at the time of rubella immunization. This would ensure infertility for the recommended 2 month post-vaccination period and would prevent needless exposure of unborn children to the defect-producing virus. (No refs.) - *N. Mize.*

2550 **HARGREAVES, T.; & PIPER, R. F.** Breast milk jaundice; effect of inhibitory breast milk and 3α,20β-pregnanediol on glucoronyl transferase. *Archives of Disease in Childhood*, 46(246): 195-198, 1971.

Breast milk from a woman whose infant showed breast milk jaundice was found to inhibit the conjugation of bilirubin in a preparation of rat liver slices. It had been hypothesized that the inhibition was due to the presence of the steroid 3α,20β-pregnanediol in breast milk; however, the breast milk was found to inhibit glucuronyl transferase within liver cells and the steroid to inhibit the secretion of bilirubin glucuronide

from the cells. The inhibitory substance within the breast milk has yet to be determined. (12 refs.) - *E. L. Rowan.*

Area Laboratories
Exeter, Devon, England

2551 **SNYDER, RUSSELL D.** Congenital mercury poisoning. *New England Journal of Medicine,* 284(18): 1014-1016, 1971.

A male infant whose mother had ingested mercury-containing hog meat during the first trimester of pregnancy was born with an elevated mercury level and developed neurologic signs attributed to mercury poisoning. The mother's urinary mercury level was not elevated. In the child, mercury levels were markedly elevated until 6 weeks of age. The child was irritable and had a weak, high-pitched cry. A series of electroencephalograms showed that after birth the infant progressed from a normal to a markedly disturbed epileptiform pattern. By 6 months he had developed generalized myoclonic jerks. By 8 months, he was irritable and hypotonic. Eye movements were nystagmoid, and there was no evidence of visual fixation. These data and other cases indicate that an asymptomatic woman who ingests organic mercury during pregnancy may produce a neurologically handicapped child with cerebral palsy, MR, convulsions, involuntary movements, and/or defective vision. (18 refs.) - *J. K. Wyatt.*

University of New Mexico
School of Medicine
Albuquerque, New Mexico 87106

2552 **BARLTROP, DONALD.** Lead poisoning. *Archives of Disease in Childhood,* 46(247): 233-235, 1971.

Few cases of lead poisoning in Britain are diagnosed, and the distribution of positive diagnoses is not uniform. Factors which make identification of poisoned children difficult include absence of specific symptoms and lack of an agreed definition of poisoning. Lead poisoning has been associated with several metabolic disturbances and encephalopathy. Suspected cases should be quickly diagnosed to avoid delay in treatment. Among the diagnostic tests are those which detect lead in the blood, those which reveal metabolic disturbances related to lead poisoning, and nonspecific tests. Treatment should consist of removal of child from source of poisoning, use of chelating drugs to remove soft-tissue lead, and prevention of recurrence. Prevention should include screening programs designed to detect and remove lead in domestic environments. (24 refs.) - *J. K. Wyatt.*

St. Mary's Hospital
London W. 2, England

2553 **CHESSELLS, J. M.; & WIGGLESWORTH, J. S.** Coagulation studies in severe birth asphyxia. *Archives of Disease in Childhood,* 46(247): 253-256, 1971.

Laboratory studies on 9 infants who were so severely asphyxiated at birth that they required mechanical ventilation for at least 10 minutes revealed disordered hemostasis in 4, 2 of whom died. Of the 4 infants in this series who died, 2 had hyaline membrane disease. None of the infants evidenced bleeding tendencies in life. There was evidence of disseminated intravascular coagulation which had occurred during the first few hours of life in 2 fatal cases and 1 survivor. Fibrin thromboemboli were present in the liver and adrenals of 2 fatal cases. Low level of vitamin K was found in 1 survivor. Although hemostatic failure related to disseminated intravascular coagulation develops in only a small minority of birth asphyxiated infants, when an infant becomes hypothermic within a few hours after birth, this possibility should be investigated. (11 refs.) - *J. K. Wyatt.*

University of Sussex
Applied Sciences Laboratory
Falmer, Brighton, England

2554 **EPSTEIN, SAMUEL S.; & ROHRBORN, GUNTER.** Recommended procedures for testing genetic hazards from chemicals, based on the induction of dominant lethal mutations in mammals. *Nature,* 230(5294): 459-460, 1971.

Testing for mutagenicity can practically and economically be done as part of routine toxicology testing *in vivo* in mammals. Male mice and rats

may receive subtoxic concentrations of a chemical and be mated with untreated females. The pregnant female can subsequently be assayed for induction of dominant lethal mutations as indicated by early fetal deaths and preimplanation losses. Acute, subacute, and chronic toxicity can be assayed by serial matings. Mutagenicity must be determined before human exposure is undertaken. (22 refs.) - *E. L. Rowan.*

Harvard Medical School
Boston, Massachusetts

2555 **WAGNER, ARNOLD L.; & McELIN, THOMAS W.** Successful pregnancy following artificial hyperimmunization to hr′(c). *American Journal of Obstetrics and Gynecology*, 110(5): 739, 1971.

A pregnancy with successful outcome was reported for an Rh-positive woman who had become severely sensitized to the hr′(c) antigen in her first marriage, volunteered to be injected with hr′(c) positive cells to maintain an antibody titer when she believed further pregnancy to be unlikely, and later desired pregnancy in a subsequent marriage to a man carrying the hr′(c) antigen. On her initial visit for this pregnancy, the indirect Coombs titer was 1:16, which rose gradually to 1:2,048. Her second husband's blood was hr′(c) positive, genotype cde/cde. When the fetus had an estimated gestational age of 34 weeks, the indirect Coombs titer remained at 1:2,048. The cervix was tightly closed, and fetal activity was minimal. The patient was delivered by caesarean section followed by bilateral partial salpingectomy. The infant weighed 2 lbs and 4 oz and had an initial cord blood bilirubin of 8.8 mg% and an initial cord blood hemoglobin of 3.4g%, with an Apgar score of 4. After 50 days of hospitalization, 4 total exchange transfusions, and several simple transfusions, the infant left the hospital. It is currently in excellent health and exhibits normal mental and physical development. (2 refs.) - *B. J. Grylack.*

2650 Ridge Road
Chicago, Illinois 60201

2556 **WILLOUGHBY, H. W.; WILLIS, T.; LEE, T. K.; FUCHIGAMI, S.; KAZEEF, M. T.; & DESJARDINS, P. D.** Study of bilirubin in amniotic fluid of rats with obstructive jaundice. *American Journal of Obstetrics and Gynecology*, 110(5): 663-666, 1971.

Two-hundred-gram female Wistar rats were investigated for the possible occurrence of maternal hyperbilirubinemia with a raised bilirubin content of amniotic fluid. Accurate monitoring of the 23-day rat gestation period was achieved, and hepatic duct ligation was carried out on control nonpregnant rats and on experimental pregnant rats on the eighteenth day of gestation. Blood and amniotic fluid were then obtained from experimental and control pregnant rats on the twenty-first day of gestation, and blood was also taken from nonpregnant controls following the operation. Both direct- and indirect-acting bilirubin were found to increase in the 3 days following hepatic duct ligation. Serum values from pregnant rats with hepatic duct ligation were much higher than those seen in controls but only up to half as high as the values seen in nonpregnant rats with hepatic duct ligation. Values for amniotic fluid samples were markedly higher for pregnant rats with hepatic duct ligation than for controls. Thin-layer chromatography of the diazotized derivatives of conjugated and unconjugated bilirubin showed that most of the material present in the extract from the test pool of amniotic fluid was unconjugated bilirubin having an R_f of 0.63 identical with the R_f for the unconjugated bilirubin standard. The data indicate that obstructive jaundice in the pregnant rat is accompanied by an increase of total bilirubin in amniotic fluid. (12 refs.) - *B. J. Grylack.*

Royal Victoria Hospital
Montreal, Quebec, Canada

2557 **DUHRING, JOHN L.; & ZWIREK, STEPHEN J.** Intrauterine fetal transfusion: Five years of experience. *American Journal of Obstetrics and Gynecology, 110(5): 670-671, 1971.*

Twenty-five fetuses were subjected to intrauterine fetal transfusion 41 times during the period 1965 to 1970. Follow-up of 6 of the 12 surviving patients showed all children to be in apparently good health, with the exception of 1 MR child who had a cardiac arrest during the second neonatal exhange transfusion and now has seizures. Of the 13 infants who died, mortality in

at least 10 was related temporally to the transfusion itself. Six were lost within 48 hours of transfusion, indicating a minimum 14.6% risk of death per transfusion. (2 refs.) - *B. J. Grylack.*

University of Kentucky Medical Center
Lexington, Kentucky

2558 **CASSADY, GEORGE; BARNETT, ROBERT; & CEBALLOS, RICARDO.** Dangers of fetal transfusion: Importance of placental location. *American Journal of Obstetrics and Gynecology,* 110(5): 672-673, 1971.

Experience to January, 1971, with 46 fetal transfusions in 28 patients with Zone III amniotic fluid suggests that the risks of transfusion may be considerably greater with anterior location of the placenta and that accurate and precise localization of the placenta is essential prior to the decision to transfuse. Nine of 28 infants represented the second pregnancy for the mother. For 14 patients, the baby requiring transfusion represented the first affected pregnancy, and 22 or 28 had not experienced a previous hemolytic perinatal death. The finding that 18 or 28 patients required initial fetal transfusion in the period August to November suggests a striking, unexplained seasonal incidence. Most perinatal losses (all 6 neonatal deaths and 9 of 13 fetal deaths) resulted from the inability to reverse fetal hydrops, which was found at initial transfusion in 16 patients. The 4 fetal deaths in nonhydropic fetuses were related to blind traversal of an anterior placenta and the ensuing laceration of a major fetoplacental vascular channel. (No refs.) - *B. J. Grylack.*

University of Alabama Medical Center
Birmingham, Alabama

2559 **SCHMID, RUDI.** More light on neonatal hyperbilirubinemia? *New England Journal of Medicine,* 285(9): 520-522, 1971.

A promising approach to the treatment of neonatal hyperbilirubinemia has been based upon the observation that bilirubin is highly photolabile. When exposed to strong illumination, particularly from a light source with peak emission in the 420 to 475 nm region, the yellow, lipid-soluble pigment undergoes gradual decomposition to a series of derivatives that exhibit progressively less yellow color and diazo reactivity but increased water solubility. Though rapidly gaining popularity, phototherapy of icteric infants has remained controversial, primarily because of concern about possible adverse effects of prolonged intense illumination and also because of uncertainty over the exact chemical nature and potential toxicity of the photochemical products of bilirubin. Evidence from available clinical studies indicates that the water-soluble pigment derivatives are not retained in the body but are excreted rapidly in bile and urine. However, some animal experiments have shown that intense illumination may alter hepatic excretory function. (11 refs.) - *B. J. Grylack.*

No address

2560 **MENSER, MARGARET A.; & FORREST, JILL M.** Immunity to rubella. *New England Journal of Medicine,* 285(7): 412-413, 1971. (Letter)

The rate of fall of rubella antibody levels with time was studied in 100 mothers of children with congenital rubella. Rubella hemagglutination-inhibition antibody levels were found to remain elevated for as long as 20 years following infection (geometric mean, 1 in 240), while the geometric-mean titer had fallen to 1 in 111 in mothers in whom infection occurred 26 to 31 years earlier. These findings suggest that immunity to rubella infection tends to decline with time. (No refs.) - *B. J. Grylack.*

Royal Alexandra Hospital for Children
Sydney, Camperdown, Australia

2561 **FORREST, JILL M.; MENSER, MARGARET A.; & BURGESS, J. A.** High frequency of diabetes mellitus in young adults with congenital rubella. *Lancet,* 2(7720): 332-334, 1971.

Fifty young adults with congenital rubella were reviewed in 1967, and diabetes mellitus was found in 1 patient. When both conditions were found in 4 further patients, an association was suspected. Of 44 patients available for reassessment in 1970, diabetes mellitus was

diagnosed in 3 males and 2 females and latent diabetes in 2 males and 2 females. In all 9 patients with diabetes, rubella infection had occurred when the mothers were from 4 to 16 weeks pregnant. One patient was below the tenth percentile in height and weight, and another was below the tenth percentile in weight at the time of testing. All were deaf, 2 had bilateral cataracts, 2 had rubella chorioretinopathy, and 1 had pulmonary-valve stenosis. Two males had undescended testes, and 1 had congenital talipes. The glucose varied in the 9 patients. The high frequency (20%) of diabetes in this group of young adults with congenital rubella strongly suggests a causative relationship between the 2 conditions. (23 refs.) - *B. J. Grylack.*

Royal Alexandra Hospital for Children
Sydney, Australia

2562 **BROWDER, J. ALBERT.** Occipital encephalocele after probable prenatal rubella. *Lancet,* 2(7720): 383-384, 1971. (Letter)

The case of a 28-month-old girl with congenital rubella and occipital encephalocele suggests a possible association between the 2 disorders. The infant was the product of a spontaneous, vertex delivery. Two occipital soft-tissue prominences association with a 1-cm bony defect, which were noted soon after birth, were resected at 19 days of age. At 2.5 years, the child manifested delayed development with mild to moderate MR, decreased visual acuity and restriction of visual fields, a right estropia, limited speech, grand-mal epilepsy, a slight intention tremor, and slight right facial weakness. She did not walk unassisted until 18 months, and her gait remains clumsy. A rubella hemagglutination titer was positive at 1/80 dilution. The child has no history of postnatal rash or illness consistent with rubella, and she has had no rubella immunization. Although a positive titer of this level is regarded as consistent with congenital disease in this case, the child does not show any other stigma of congenital rubella in cardiac, eye, or ear function. (6 refs.) - *B. J. Grylack.*

University of New Mexico School of Medicine
Albuquerque, New Mexico 87106

2563 **GERSON, KENNETH L.; & HASLAM, ROBERT H. A.** Subtle immunologic abnormalities in four boys with subacute sclerosing panencephalitis. *New England Journal of Medicine,* 285(2): 78-82, 1971.

The immunologic capabilities in 4 boys with proven subacute sclerosing panencephalitis (SSPE) were examined in order to determine more clearly the pathogenesis of this slow virus infection. An absence of serum immunoglobulin A (IgA) was observed in 1 patient, and 2 of the remaining 3 had a deficiency of circulating IgA. Sections of regional lymph nodes obtained after booster immunization with diphtheria-tetanus toxoid showed suggestions of primary lymphoid follicles in the cortex, but germinal centers were rare and small. Delayed hypersensitivity responses to intradermal antigens showed absent or greatly decreased responses in all cases. Sensitization to dinitrochlorobenzene (DNCB) failed to produce contact sensitivity when challenged with 100 μg of DNCB 14 days later. Skin allografts were rejected but with a moderately prolonged rejection time. The obtained data indicate that a subtle functional defect in the thymus system of cells on either a congenital or an acquired basis may play a role in this disease. (28 refs.) - *B. J. Grylack.*

University of Kentucky Medical Center
Lexington, Kentucky

2564 **LAPIN, JOSEPH H.** A vaccination dilemma. *Journal of the American Medical Association,* 217(1): 80, 1971. (Letter)

Recent advice that laws requiring rubella vaccination as a condition for school entrance should be repealed or suspended until it can be determined whether they are in the best interests of the community, and that the lay public should be made aware of both the hypothetical risks and advantages of mass vaccination, has illustrated an insoluble dilemma. Although private pediatricians generally oppose mumps vaccination for anyone except adolescent boys, the same situation may arise in this disease, resulting in loss of immunity before puberty. However, the medicolegal status of the pediatrician is uncertain, and it is questionable whether he can insist, against the law, on the admission into school of children without rubella vaccination. (No refs.) - *B. J. Grylack.*

No address

2565 **BALDWIN, J. A.; & FREESTONE, D. S.** Risk of early post-partum pregnancy in context of post-partum vaccination against rubella. *Lancet*, 2(7720): 366-367, 1971.

The frequency of conception was investigated in all women residing in the Oxford, Oxfordshire (England) region who had their first liveborn child in 1963 in an attempt to estimate the possible risks of post-partum vaccination against rubella. Of 2,124 women, 700 (33%) entered second pregnancies within 2 years of first delivery. One (0.05%) and 21(1.0%) conceived within 60 and 90 days of delivery, respectively. If it is assumed that conception occurred 2 weeks after the start of the last menstrual period, that the period of risk following vaccination is approximately 40 days, and that 20% of primiparous women are seronegative, no women in the cohort who entered second pregnancies would have risked fetal damage if vaccinated within 15 days of delivery. Thereafter, the risk of fetal damage would have increased. If the period of risk is assumed to be 60 or 90 days, 0.2 and 4.2 pregnancies, respectively, would have been in danger even from immediate postpartum vaccination. In the light of these findings, the importance of the earliest possible vaccination after delivery and of avoiding conception in the 2 to 3 months postpartum cannot be overemphasized. (8 refs.) - *B. J. Grylack.*

University of Oxford
Unit of Clinical Epidemiology
Oxford, England

2566 Irradiation of transfused blood helps to prevent graft-versus-host disease. *Journal of the American Medical Association*, 217(8): 1037, 1040-1041, 1971.

Investigators at 2 universities have suggested a way to prevent development of fatal graft-versus-host (GVH) disease following a blood transfusion in patients with an immune system deficiency. The clinical and laboratory studies reported by the 2 groups advised that irradiation of the blood before transfusion destroys the ability of transfused lymphocytes to replicate and take over the host. Radiation was used for selective lymphocyte destruction, since cells capable of replication have been shown to be more radiosensitive than platelets, red cells, or granulocytes. It was also suggested that transfusions of irradiated lymphocytes may help patients with severe combined immunodeficiency to fight infection until a suitable matched donor can be found. When 1 group of investigators administered irradiated blood to 2 infants with combined immune deficiency disease, 1 infant with disseminated vaccinia resulting from a smallpox vaccination showed some evidence of improvement but died 10 days after the first transfusion and 48 hours after the second. The pneumonia affecting the second infant cleared completely, but he died of respiratory complications 12 days after reconstitution. (No refs.) - *B. J. Grylack.*

2567 **WEBB, H. E.; ILLAVIA, S. J.; & LAURENCE, GWENNETH D.** Measles-vaccine viruses in tissue-culture of non-neuronal cells of human fetal brain. *Lancet*, 2(7714): 4-5, 1971.

The growth of live measles virus vaccine strains in human fetal brain tissue-cultures was investigated. Ten tissue-culture tubes of brain cells were set up with 2 viruses, and 10 tubes were used as controls. The infected cells proliferated more rapidly than the controls. These findings are discussed with respect to the chronic virus-caused diseases of the central nervous system. (12 refs.) - *T. C. Turner.*

St. Thomas's Hospital
London, England

2568 **PAMPIGLIONE, G.; GRIFFITH, A. H.; & BRAMWELL, E. C.** Transient cerebral changes after vaccination against measles. *Lancet*, 2(7714): 5-8, 1971.

The effect of live measles vaccine on the central nervous systems of 8 experimental Ss and 3 control children is reported. All vaccinated children developed some symptoms on days 7 and 8 after vaccination, which completely disappeared by the fourteenth day. The control Ss had neither fever nor rash. Definite electroencephalogram (EEG) changes were seen in 6 of 8 experimental Ss, who had returned to normal by day 14. These findings are discussed in light of other studies in the field and the necessity for further research cited. (20 refs.) - *T. C. Turner.*

Hospital for Sick Children
London, England

2569 Can we stop the march of slow viruses? *Medical World News,* 12(April 2): 25-28, 1971.

Slow-virus diseases are prolonged degenerative disorders of the nervous system and can take several years for their symptoms to become clinically manifest. Kuru, Creutzfeldt-Jakob disease, and subacute sclerosing panencephalitis have been traced to a slow-virus agent, while multiple sclerosis and postencephalitic parkinsonism are among the suspected slow-virus diseases. It is thought by some investigators that aging itself may involve a slow-virus infection. Nothing has yet been found, however, to successfully treat the slow-virus infections. (No refs.) - *T. C. Turner.*

2570 Measles — quick and slow. *Lancet,* 2(7714): 27-28, 1971.

Two main types of "slow" neurological disease which have been discovered in man (kuru and Creutzfeldt-Jakob disease; subacute sclerosing panencephalitis (SSPE) and progressive multifocal leukoencephalopathy) are discussed, recent work on SSPE is noted, and experiments done by researchers on measles virus are reported. Propagation of a vaccine strain of measles virus in tissues of human fetal brain has revealed an abnormal degeneration-proliferation of cells characteristic of malignancy. Such behavior indicates that the virus has permanently damaged the metabolic and functional properties of the cells. More highly refined experiments are needed to determine the respective roles of neurones and glial cells, to illuminate the action of measles virus as compared to "slow viruses" in neurological diseases. Other experimenters have recorded electroencephalographic changes in 6 out of 8 children studied before and after vaccination with live measles virus; these abnormalities deserve fuller study. Since a method of preparing cells chronically infected with measles or other virus has become available, it should now be possible to reject or prove many current hypotheses regarding slow viruses and their part in causing parkinsonism and other chronic degenerative diseases. (27 refs.) - *T. C. Turner.*

2571 **BROWDY, SOL.** Should every newborn infant be given a Coombs test? *Clinical Pediatrics,* 10(7): 427-430, 1971.

The necessity and value of the Coombs test for diagnosing hemolytic disease in the newborn was evaluated by comparing the laboratory results with the clinical findings in 1,300 cord blood specimens tested because of potential Rh or ABO mother-child incompatibility. In 8.1% of the cases infants were Coombs positive. Of these 105 Coombs positive infants, 82 had no clinical signs or symptoms of hemolytic newborn disease. The term "subclinical disease" was reserved for such Coombs positive infants who did not become jaundiced in the first 48 hours of life, if at all, and whose bilirubin levels did not exceed 15 mg/100 ml. Since the 23 Coombs positive infants with clinical hemolytic disease would have come to attention by the second day of life because of their jaundice, even without the cord examinations, the hoped-for advantages of early identification with the Coombs test are seen to be outweighed by the disadvantages of overburdening the staff, financial cost, and needless arousal of anxiety. While a Coombs test on all infants of Rh negative mothers is recommended, routine testing of cord blood for potentially ABO cases of hemolytic disease is not recommended. (4 refs.) - *K. L. Woodworth.*

446 Bellevue Ave.
Trenton, New Jersey 08618

2572 **WELLER, THOMAS H.** The cytomegaloviruses: Ubiquitous agents with protean clinical manifestations (first of two parts). *New England Journal of Medicine,* 285(4): 203-214, 1971.

The progress report on the cytomegaloviruses (CMV), members of the herpesvirus family, emphasizes the clinical and epidemiologic importance of the human CMV. Since an immunologic approach to differentiation of the herpesvirus family should be more productive than a biologic one, pertinent virologic and immunologic attributes of the CMV are summarized. A brief survey of the CMV in animals precedes review of the virologic and immunologic attributes of the CMV in man: characteristics of the CMV under conditions of *in vitro* cultivation, viral assay techniques (particularly plaque assay), and serologic procedures for detecting antibodies produced in response to infection with a CMV (neutralization, complement-fixation, immunofluorescence, and platelet-agglutination). Information is also presented on pathogenesis at the

cellular level and on host-parasite interactions. Virus-cell interactions are examined in terms of susceptibility to viral replication at the cellular level, capacity of CMV to induce interferon and sensitivity of CMV to it, concomitant cellular infection with a CMV and with toxoplasma or pneumocystis, and cellular alteration or transformation. Discussion of host-parasite interactions covers new pathogenic concepts about CMV acquired postnatally and during pregnancy, immunologic aberrations, occult viremia (cytomegaloviremia), and interactions in the abnormal host (patients with generalized neoplasia and the renal allograft recipient). (127 refs). - *K. L. Woodworth.*

Harvard School of Public Health
Boston, Massachusetts 02115

2573 **KATZ, MICHAEL.** Viruses in brain cells. *Lancet,* 2(7721): 437-438, 1971 (Letter)

In brain-cell cultures from patients with subacute sclerosing panencephalitis (SSPE), the cells, although they did not yield infectious virus directly, showed changes in their growth pattern, characterized by proliferative activity and loss of contact inhibition. In addition, their karyotypes became aneuploid. Animal experiments with SSPE viruses and rabies virus showed that EEG abnormalities may signal presence of virus in the brain. Abnormal EEG patterns allowed identification of ferrets (injected with brain homogenates of SSPE patients, or with tissue-culture cells infected with SSPE viruses) who developed encephalitis but exhibited few or no clinical symptoms. In the same manner, abnormal EEG patterns detected encephalitis in rats infected with rabies virus 3 days before there was clinical evidence of encephalitis. (4 refs.) - *K. L. Woodworth.*

Columbia University School of
Public Health and Administration Medicine
New York, New York 10032

2574 **MIALE, JOHN B.** Rh immunization. *Journal of the American Medical Association,* 217(3): 346, 1971.

The RH Problem, edited by John G. Robertson and Francesco Dambrosio (Milan, Italy, Instituti Clinici de Perfezionamento, 1970, 242p. $10.00), is recommended for readers seeking advanced information on Rh immunization. Containing the proceedings of an International Symposium on the Prevention and Treatment of Rh Immunization, the book concerns prevention of Rh immunization, antepartum assessment of patients with Rh immunization, intrauterine treatment of the fetus, and various special problems as they relate to Rh hemolytic disease of the newborn. Each contributor is an authority in his field and covers his topic in great detail. (No refs.) - *K. L. Woodworth.*

University of Miami
Miami, Florida

2575 **BELLANTI, JOSEPH A.; CATALANO, LOUIS W.; & CHAMBERS, ROBERT W.** Herpes simplex encephalitis: Virologic and serologic study of a patient treated with an interferon inducer. *Journal of Pediatrics,* 78(1): 136-145, 1971.

This case history of a 4½-year-old Caucasian boy, the first recorded description of treatment of a human with the interferon inducer In.Cn, and the first documented interferon study of herpes-simplex virus encephalitis in man, does not establish a definite therapeutic effect, although S apparently improved clinically while receiving the drug. Elevated blood urea nitrogen and other parameters after 11 days of treatment suggested mild toxicity. High interferon levels before drug administration prevented establishing that interferon induction by In.Cn had occurred, and indicated that severe herpes-simplex infection strongly stimulates interferon production. (40 refs.) - *B. Berman.*

Georgetown University School of Medicine
Washington, D. C.

2576 **SCHIFF, D.; CHAN, G.; & STERN, L.** Fixed drug combinations and the displacement of bilirubin from albumin. *Pediatrics,* 48(1): 139-141, 1971.

During investigation of the effects of bilirubin binding of certain drugs used in the neonatal period, the effects of fixed drug combinations on the displacement of bilirubin from albumin were assessed in caffeine sodium benzoate and

diazepam. Sephadex G-25 gel filtration and bilirubin spectral curves were used for the evaluation. The fixed combination of caffeine sodium benzoate in recommended therapeutic doses (64 mg/ml) was found to affect the binding by both methods. Sodium benzoate alone was shown to uncouple bilirubin from the albumin complex. Diazepam in its injectable form appeared to be a potent bilirubin-albumin uncoupler. The data have demonstrated that the therapeutically active component of the combination, namely caffeine and diazepam, is not involved in the competition with bilirubin for the albumin binding sites, but the preservative component, sodium benzoate, is. In assessment of the displacement properties of any drug used in the neonatal period, it is important that the various components of the preparation be evaluated both individually and in combination. (7 refs.) - *B. J. Grylack.*

Montreal Children's Hospital Research Institute
Montreal 108, Quebec, Canada

2577 SEVER, JOHN L. Rubella immunization risk postpartum. *Journal of the American Medical Association*, 217(5): 697, 1971. (Letter)

Data on 53,000 pregnant women at 12 United States medical centers were studied in order to evaluate policy concerned with the risk of rubella immunization in postpartum women. It was determined that 5.6% of these women became pregnant within 2 months following delivery. Since there is at least a theoretical risk to the fetus in vaccination of pregnant women, medically accepted methods for pregnancy prevention should be followed carefully when rubella immunization is given to postpartum women. (3 refs.) - *B. J. Grylack.*

No address

2578 NOBLE, JAMES S.; & WAND, MARTIN. Catcher's crouch and rubella immunization. *Journal of the American Medical Association*, 217(2): 212, 1971. (Letter)

Two cases of the "catcher's crouch syndrome" were observed in boys aged 6.5 years following rubella vaccinations with intervals of 47 and 41 days, respectively. In both children, onset was marked by severe bilateral knee pain and inability for leg extension, which caused the boys to remain in crouched positions. Aspirin was the basic treatment. The time range for complete resolution varied from 4.5 months in 1 case to 3.5 weeks in the other. These symptoms have been reported elsewhere and probably represent only 1 facet of a spectrum of symptoms following rubella vaccination. Rheumatic fever, juvenile rheumatoid arthritis, and other entities should be ruled out with these symptoms. Despite the fact that manifestations of the symptom complex of paresthesia and joint pain have become linked more commonly to rubella vaccination as time progresses, these benign complications should not detract from continuation of rubella vaccination programs. (No refs.) - *B. J. Grylack.*

2579 FIERZ, L. Immunological aspects of the Guillain-Barre syndrome. *Lancet*, 2(7717): 211, 1971. (Letter)

Necropsy findings of widespread CNS involvement in the Guillain-Barre syndrome fit in well with the presence of encephalitogenic-factor-sensitized lymphocytes. CNS damage in the presence of anatomical lesions may not be clinically evident. In severe peripheral motor and sensory disturbance, classic CNS signs do not help and mental symptoms may be difficult to evaluate. (1 ref.) - *B. Berman.*

Clinique Universitaire de Medecine
1200 Geneva, Switzerland

2580 WELLER, THOMAS H. The cytomegaloviruses: Ubiquitous agents with protean clinical manifestations. *New England Journal of Medicine*. 285(5): 267-273, 1971.

Surveys show that risk of acquired infection with cytomegaloviruses (CMV) begins at birth (revealed by viruria), with cumulative risk during life. CMV infections can be acquired through congenital infection, prenatal iatrogenic transmission, during birth via cervical secretions, or postnatally. The immature nervous system is a frequent target (CMV may be responsible for focal infections); low birth weight may indicate viruria. Postnatally, CMV may affect liver, spleen,

hematopoietic system, kidneys and lungs; the nervous system is spared in children and adults. Diagnosis is best made by demonstrating viruria through inoculation of appropriate tissue cultures. Therapy has included use of interferon, and topical use of inhibitors of DNA viruses in treating systemic herpes-group infections. Endemic in all human societies, CMV, with their broad, serious illness consequences, pose large social problems. (184 refs.) - *B. Berman.*

Harvard School of Public Health
Boston, Massachusetts

2581 **GRIFFITH, JOHN F.** Nervous system manifestations of cytomegalovirus (CMV) infection. *Developmental Medicine and Child Neurology*, 13(4): 520-522, 1971.

The nervous system is involved in about 10% of cases of generalized cytomegalovirus (CMV) infection, with manifestations variable both clinically and pathologically. In severely affected infants, their brains usually show a diffuse necrotizing encephalitis with perivascular lymphocytic infiltration in parenchyma and meninges, microglial nodules, nerve cell loss, hemorrhage and, rarely, cytomegaly. Although the evidence implicating CMV in the pathogenesis of congenital anomalies of the central nervous system is slight, there is no question that neural tissue can be injured by the encephalitis process, leading to functional and morphologic alterations. There is now no evidence suggesting that adults who become infected with CMV develop nervous system infection or experience chronic neurologic disability. However, there is reason for concern about nervous system function in infants with asymptomatic CMV infection, since reports have indicated that a small but significant percentage of such infants do ultimately demonstrate neurologic dysfunction (microcephaly and MR in particular). More data are required concerning whether neurologic sequelae develop a significant percentage of patients with persistent asymptomatic CMV infection. (8 refs.) - *K. L. Woodworth.*

Duke University Medical Center
Durham, North Carolina 27710

2582 **SCHAIN, RICHARD J.; & WILSON, GABRIEL.** Brainstem encephalitis with radiographic evidence of medullary enlargement. *Neurology*, 21(5): 537-539, 1971.

Radiotherapy or surgical exploration in cases of brainstem encephalitis is best deferred for 8 weeks after first symptoms so as to avoid diagnostic confusion with encephalitic syndromes. This is illustrated in the case of a 3-year-old child who one week after recovery from a flu-like illness displayed multiple bilateral cranial nerve palsies, gait disturbance, and fine-motor incoordinations. Medullary glioma was indicated in radiographic studies. Confirmation of brainstem diagnosis came when all neurologic symptoms disappeared after 5 weeks. In 18 months, S was completely normal. Absence of symptoms at this interval ruled out brainstem glioma. (8 refs.) - *B. Berman.*

UCLA Center for the Health Sciences
Los Angeles, California

2583 **AMATO, R. S.; & PELLEGRINO, R. J.** Chromosomes of cells cultured from brain tissue of a patient with subacute sclerosing panencephalitis. *Neurology*, 21(5): 553-558, 1971.

A culture of brain tissue from a 10-year-old boy with subacute sclerosing panencephalitis (a disease associated with measles virus in the CNS) showed instability of chromosomal number (elevation occurred in 21% of harvested cells) and morphology. The increased number in the first transfer were generally in the 80-90 chromosome range; 7% of these were endoreduplications. The modal number varied from hypotetraploid to hypertetraploid. Elevated chromosome numbers (60-70 range) as well as breaks, exchanges, fragments, and marker chromosomes were seen by the seventh transfer. S, admitted for progressive dementia, had gone through physical, mental, and personality changes, and is currently hospitalized. (19 refs.) - *B. Berman.*

University of Nebraska College of Medicine
Omaha, Nebraska

2584 **EMANUAL, IRVIN; & ANSELL, JULIAN S.** Congenital abnormalities after mumps vaccination in pregnancy. *Lancet*, 2(7716): 156-157, 1971. (Letter)

A mother who was given live mumps vaccine 17 days after beginning of her last menstrual period at estimated 2-4 days post coitus bore a child with exstrophy of the urinary bladder. Other defects were deformed genitalia, duplication of descending colon with two rectal openings, recto-vaginal fistula, and hemivertebrae T7 to T11. Both parents were in good health, already had a normal boy, and there was no family history of congenital malformations. Exstrophy of the bladder is a rare anomaly (0.05 cases per 1,000 in one study). Little is known of the teratogenic properties of wild mumps virus and reports are conflicting. This case may be considered a chance association, but extreme care should be taken in administering live virus vaccines of unknown teratogenic potential to women during childbearing years. (9 refs.) - *S. M. Rucker.*

University of Washington
Seattle, Washington 98105

2585 **MacMAHON, BRIAN; & YEN, STELLA.** Influenza and neural-tube defects. *Lancet*, 2(7718): 260, 1971. (Letter)

Statistical analysis of available data does not support a suggestion by Janerich that the epidemic of neural-tube defects which extended over a period of 25 yrs and peaked in the early 1930s may have been caused by exposure of prepubescent girls to influenza in the 1918 epidemic. If the hypothesis were valid, a shift in the maternal-age distribution of affected births should have occurred during the period of the epidemic (as Janerich noted). Available data indicates a nearly constant mean maternal age (MMA) at birth during the epidemic whereas the predicted mean age, assuming that the epidemic component was contributed by a cohort of women who were about 12 yrs old in 1918, increased by 5 yrs over the 25-year period. A possible explanation considered was that the maternal age shift occurred but was offset by a general lowering of the MMA in the population of births at risk. However, data on unaffected births from two hospitals from which the series was assembled also showed an almost constant mean maternal age at birth during the 25-year period. These data rule out any cohort pattern phenomenon as an explanation of the epidemic. Existing knowledge of the causes of congenital malformations would suggest changes in factors operative in early pregnancy as a likely explanation, but the specific change involved in this episode is still unknown. (2 refs.) - *S. M. Rucker.*

Harvard School of Public Health
Boston, Massachusetts 02115

2586 **ROONEY, JOHN C.; HILL, DAVID J.; & *DANKS, DAVID M.** Jaundice associated with bacterial infection in the newborn. *American Journal of Diseases of Children*, 122(1): 39-41, 1971.

Bacterial infection should be considered as a possible cause of neonatal jaundice whether the baby is well or ill. In fact, jaundice can be the only obvious symptom of bacterial infection in newborn babies. The findings are based on the clinical records of 22 Ss selected from the files of the Royal Children's Hospital, Melbourne, for the period July 1963 to December 1969. The criteria for selection were jaundice of abnormal intensity or duration, definite proof of a bacterial infection, and adequate exclusion of other known causes of jaundice. Nine babies were active and well throughout, with jaundice the only abnormal clinical finding. Six were lethargic and slow eaters but otherwise well. Seven were ill with vomiting, dehydration, or other symptoms. There were no deaths in the series. All patients recovered uneventfully and only one, a child with clinical evidence of kernicterus, was abnormal at discharge. The jaundice cleared within 6-7 days after beginning antibiotic therapy. In 11 Ss, positive blood culture was the only indication of infection, and *E. coli* was isolated in 6 of these. Nine had urinary tract infections, one had purulent meningitis, and one had periumbilical cellulitis. Leukocytosis was found in only 9 Ss and anemia in only one. There were 19 boys and 3 girls in the sample, of which 5 were born prematurely. The principal finding is that bacterial infection can be associated with jaundice in babies who are not ill. Blood culture and urinary examination should be performed in all newborn babies with jaundice for which there is no obvious cause. (7 refs.) - *S. M. Rucker.*

*Royal Children's Research Foundation
Victoria, Australia

2587 **EICHHORN, MARY M.** Rubella: Will vaccination prevent birth defects? *Science*, 173(3998): 710-711, 1971.

The value of rubella vaccine in preventing congenital rubella is discussed. It is not certain whether the vaccines can safely achieve the goal of protection of pregnant women. The U.S. Public Health Service (PHS) recommends that all young children be vaccinated and suggests vaccination of susceptible, nonpregnant women on an individual basis. Some scientists disagree, advocating that susceptible women and adolescent girls be the primary candidates for vaccination, since vaccination of small children may lead to a future population of young women who would be more susceptible than if there had been no vaccine, because trials have indicated that vaccine does not give as complete immunity as the disease does. Several strains of rubella virus have been developed and tested; some are quite effective, but some cause unpleasant side effects, such as temporary arthritis. Another factor important in determining the value of a vaccine is the immunologic response. None of the vaccines produce as many antibodies in the blood as natural infection. Clinical trials of vaccine protection (of men at Ft. Ord, California, and of institutionalized children) report conflicting results: 80 percent of the vaccinated men were reinfected and only 3.4 percent of the naturally immune men; 50 percent of the children were reinfected, but neither group became ill. At a boarding school, 5 of 22 became infected. There is a danger that a reinfected vaccinated person may transmit a wild-type virus to a susceptible person. Authorities agree that several years of experience with the vaccine will be necessary before its value in preventing congenital rubella is known. (4 refs.) - *T. C. Turner.*

2588 **HORTA-BARBOSA, LUIZ; HAMILTON, REBECCA; WITTIG, BARBARA; FUCCILLO, DAVID A.; SEVER, JOHN L.; & VERNON, MINA LEE.** Subacute sclerosing panencephalitis: Isolation of suppressed measles virus from lymph node biopsies. *Science,* 173(3999): 840-841, 1971.

The fact that subacute sclerosing panencephalitis (SSPE) is not limited to the central nervous system is demonstrated, and a suggestion made that it may be associated with a specific defect of cellular immunity. Lymph node biopsies were taken from 5 Ss with SSPE and 5 controls. A part of each node was washed in Hanks balanced salt solution and minced into fragments which were suspended in Eagle's minimum essential medium (EMEM) enriched with 20 percent fetal bovine serum. After 3 days cultures showed fibroblasts growing from edge of explants and nonattaching cells. Both types of cells showed normal morphology when fixed and stained with eosin and hematoxylin. Nonattached cells were decanted, centrifuged, and resuspended in Roswell Park Memorial Institute culture medium. This was used to prepare mixed cultures with the Ohio strain of Hela cells, which were incubated at 37°C. A table gives the lists of isolations obtained from the patients in the study (3 negative, 2 positive). Measles virus was isolated from the lymph node cells of SSPE patients 3 and 5. Tissue from each node was put under electron microscopy and intracytoplasmic filamentous structures were seen in the node tissue of SSPE patient 3, but this could not be considered conclusive evidence of myxoviruses, since reticular fibers may have similar configurations. The 2 isolations came from patients with early clinical symptoms of the disease, while measles virus could not be isolated from patients in later stages of SSPE. It is hypothesized that a chronic intracellular viremia persists during the SSPE incubation period. These measles-carrier cells would move to the lymph node and eventually to the central nervous system to begin the neurological phase of SSPE. SSPE virus infection is not restricted to the brain alone. (8 refs.) - *T. C. Turner.*

National Institute of Neurological
Diseases and Stroke
Bethesda, Maryland 20014

2589 **POSKITT, E. M. E.** Effect of measles on plasma-albumin levels in Ugandan village children. *Lancet,* 2(7715): 68-70, 1971.

In light of the finding that 24% of the 150 children most recently admitted with kwashiorkor or marasmus to the Child Nutrition Unit in Kampala (Uganda) had measles 2 months or less before admission, the possible implication of measles in the development of kwashiorkor was investigated. The Ss, younger than 3 years of age, were cases of measles attending a weekly rural clinic. The mean albumin levels and S.E. 4 to 6 weeks before measles in individuals bled before and during measles was 3.34 ± 0.12 g per 100 ml. During the measles episode, the corresponding value was 2.98±0.09 g per 100 ml. This difference was statistically significant ($t=3.36$, $p<0.005$). The mean albumin level during

measles in the group bled at the time of and 6 weeks after measles was 3.00±0.07 g per 100 ml, but during recovery this value had risen to 3.42±0.11 g per 100 ml. This difference was also significant ($t=4.05$, $p<0.001$). Inpatients who developed measles during treatment for kwashiorkor also showed a fall in plasma-albumin concentration despite a diet that was adequate in calorie and protein intake for the growth and regeneration of albumin. Although measles alters the apparent nutritional state even under conditions of good diet, the same good diet produces a rapid rise of plasma-albumin concentration in convalescence. (5 refs.) - *B. J. Grylack.*

Medical Research Council
Kampala, Uganda

2590 Coagulation defects in the newborn. *Lancet,* 2(7715): 85, 1971.

Despite methodological advances, the etiology and pathogenesis of intracranial hemorrhage, hemorrhage in other organs, and the formation of fibrin-rich membranes in infantile respiratory distress syndrome remain conjectural. Micromethods have been developed for estimating various clotting factors on capillary blood samples which would facilitate closer monitoring in small infants. However, research has demonstrated that the sera of hypoxic guinea pigs and hypoxic newborn babies contain humoral anticoagulants. Evidence appears to indicate that thrombotest is not a useful screening test for babies with a hemorrhagic tendency who are at risk from intracranial bleeding, in contrast to a previous report. Although pathological data from necropsies show that intravascular microthombi are commonly found in association with pulmonary hyaline membranes, the clinical significance of occasional or moderate microthrombi formation remains difficult to assess. Despite encouraging results from experimental administration of heparin to children with meningococcemia, the efficacy of the drug is still uncertain, since the trial was not controlled. (No refs.) - *B. J. Grylack.*

2591 **DODGE, J. S.** Rubella vaccination. *Lancet,* 2(7715): 93-94, 1971. (Letter)

Epidemiologically, it appears wise to offer immunization against rubella first to children of both sexes aged 5 to 10 years. Besides protecting the girls for the future and greatly reducing the number of susceptibles who are most involved in transmitting rubella, this immunization would delay considerably the onset of the next rubella epidemic. (1 ref.) - *B. J. Grylack.*

University of Otago
Dunedin, New Zealand

2592 **LOPEZ, C. G.; & LIE-INJO LUAN ENG.** Alpha-thalassaemia in newborns in West Malaysia. *Human Heredity,* 21(2): 185-191, 1971.

In an attempt to distinguish different α-thalassemia genotypes in West Malaysia, 1,015 randomly collected samples of cord blood from 568 Chinese, 205 Malay, and 226 Indian babies taken immediately after delivery were studied by starch-gel electrophoresis, and quantitative studies of hemoglobin (Hb) Bart's were done when it was present. Results showed 7.0% of the Chinese, 4.9% of the Malay, and 1.8% of the Indians with Hb Bart's. Quantitation of Hb Bart's gave results ranging from 1.6 to 12.3% to 2.9% in the 4 affected Indians. As expected, the incidence of Hb Bart's was highest in Chinese infants and lowest in Indian infants. The slightly higher figures in this series as compared with previous studies were probably due to the more sensitive medium of starch-gel electrophoresis. In analysis of the Hb Bart's component, 2 distinct categories were found, 1 ranging from 1 to 3.2% and the other from 5.1 to 12.3%. This differentiation suggested the existence of 2 genotypes that had been designated recently as $\alpha\text{-thal}_1$ and $\alpha\text{-thal}_2$. (22 refs) - *B. J. Grylack.*

Institute for Medical Research
Kuala Lumpur, Malaysia

2593 **EDSALL, GEOFFREY.** Experiments at Willowbrook. *Lancet,* 2(7715): 95, 1971. (Letter)

Krugman and Giles were justified in inducing hepatitis artificially in MR children who were patients at the Willowbrook State School. Aside from their own explanation that natural hepatitis infection occurs sooner or later in virtually 100% of patients admitted to Willowbrook, it seems to

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be proper and ethical to carry out experiments in children which would apparently produce no greater risk than the children are likely to incur by nature and from which the parents as well as the physician feel that a significant contribution to the future well-being of similar children may result. (2 refs.) - *B. J. Grylack.*

State Laboratory Institute
Boston, Massachusetts 02130

2594 **LOWENTHAL, A.; VAN SANDE, M.; & KARCHER, D.** Serum gamma globulins in 84 typical cases of subacute sclerosing panencephalitis. *Neurology,* 21(3): 277-280, 1971.

A total of 115 serum specimens was collected from 84 patients in order to study serum gamma globulins in typical cases of subacute sclerosing panencephalitis (SSPE). Virus neutralization, immunofluorescence, and complement fixation were used to demonstrate antibodies. Agar electrophoresis revealed gamma globulin content, and the relative mobility of the M gradients was calculated in function of the mobility of human serum albumin and dextran migrating on the same electrophoretic slide. A significant increase in SSPE serum gamma globulin relative concentration was shown. In 79 of the 115 serum samples, well-prounounced M gradients were visualized in the electrophoretic pattern of the gamma globulin. Their presence in the sera underlines the existence of a protein metabolic disorder. This gamma globulin pattern differentiates SSPE from measles and myeloma. The basic immunologic process is still poorly defined. (19 refs.) - *B. J. Grylack.*

Born-Bunge Foundation for Research
Bechem-Antwerp, Belgium

2595 **HEGGIE, ALFRED D.** Pathogenesis of the rubella exanthem: Isolation of rubella virus from the skin. *New England Journal of Medicine,* 285(12): 664-666, 1971.

To determine whether rubella virus is present in the skin at sites involved by the rubella exanthem, throat gargles, serum samples (immediate and 3 weeks later), and a 3-mm punch biopsy of a rubella maculopapule were obtained from 14 young men seen within 24 hours after appearance of the rash. Throat garglings and comparison of serum pairs established diagnosis of rubella. Rubella virus was recovered from the fluids of 13 of the 14 skin cell cultures from the rubella Ss, and from none of the skin cultures from 12 control Ss. Detection of virus in only 1 of the 14 first serum specimens makes serum an improbable source of virus in these cultures. Inoculation with rubella virus of skin cell cultures from three controls caused replication of the virus in the inoculated cultures, despite the controls' serum antibody titers indicative of previously acquired immunity. The virus was also recovered from skin cells after 6 months in culture and from skin fibroblast subcultures after as many as 7 passages, indicating that these cells are persistently infected and can be regarded as carrier cultures. Although the study shows that infectious rubella virus is present in the erythematous maculopapular skin lesions associated with rubella, the explicit role of virus in the production of rash remains unresolved. (11 refs.) - *K. L. Woodworth.*

Naval Medical Research Unit No. 4
Great Lakes, Illinois 60088

2596 **MICHAELS, RICHARD H.** Brief recording: Increase in influenzal meningitis. *New England Journal of Medicine,* 285(12): 666-667, 1971.

During the past 25 years, *Haemophilus influenzae* meningitis admissions to the Children's Hospital of Pittsburgh have increased by 400%, while total admissions have increased only 50%. Admissions for meningitis due to *Diplococcus pneumoniae* or *Neisseria meningitidis* have not increased. No evidence has been found that the greater numbers of admissions for influenzal meningitis are due to increased referral to the hospital from a wider geographic area, improved laboratory methods for detection, or change in age distribution which might reflect age-specific levels of immunity and placentally transferred maternal antibodies. A similar trend in increase of influenzal meningitis has been reported elsewhere in the United States, as well as in several foreign countries. It would be useful if others would document their experience with influenzal meningitis or suggest other possible explanations for the apparent increase. (11 refs.) - *K. L. Woodworth.*

Children's Hospital of Pittsburgh
Pittsburgh, Pennsylvania 15213

2597 **CLARK, JOHN L.** Detection of lead toxicity. *New England Journal of Medicine*, 285(11): 636, 1971. (Letter)

In an article (*N Engl J Med* 284: 565-569, 1971) on correlation between δ-aminolevulinic acid dehydratase(ALAD) activity and blood lead levels, the regression line (Fig. 1) seems to be misplaced. The regression of blood lead upon the log of ALAD activity must be intended, since the article suggests the use of ALAD assays as a screening test instead of blood lead determination. A regression equation was determined after estimation of the data points from the scatter diagram. This equation is quite different in both slope and intercept from a regression equation for blood lead upon the log of ALAD activity derived from data presented by Hernberg et al. (*Arch Environ Health*, Chicago 21:140-145, 1970). Some other possible curves using the estimated data points were fitted. Others might be tried too, but the point is that the proper mathematical form to express the relation between ALAD activity and blood lead levels is yet to be determined. More work must precede the confident use of ALAD assays in place of blood lead determinations. (2 refs.) - *K. L. Woodworth.*

U.S. Public Health Service
Bureau of Community Environmental Management
Cincinnati, Ohio

2598 **OSKI, FRANK A.** Detection of lead toxicity. *New England Journal of Medicine*, 285(11): 636, 1971. (Letter)

The regression line in Figure 1 in our article (*N Engl J Med* 284: 565-569, 1971) on the correlation between δ-aminolevulinic acid dehydratase (ALAD) activity and blood lead levels was calculated on the ALAD values up to a blood lead of 100 μg. There was apparent lack of sensitivity beyond this point. A red-cell ALAD assay of less than 15 units indicates blood lead in excess of 60 micrograms per cent and an increased body lead burden. This ALAD value is similar to that observed by Hernberg et al. (*Arch Environ Health*, Chicago 21: 140-145, 1970) for a blood lead level of 60 μg. (No refs.) - *K. L. Woodworth.*

Children's Hospital of Philadelphia
Philadelphia, Pennsylvania

2599 **NOVY, MILES J.; FRIGOLETTO, FREDERIC D.; EASTERDAY, CHARLES L.; UMANSKY, IRVING; & NELSON, NICHOLAS M.** Changes in umbilical-cord blood oxygen affinity after intrauterine transfusions for erythroblastosis. *New England Journal of Medicine*, 285(11): 589-595, 1971.

The oxygen affinity of umbilical-cord blood was measured in 15 infants with erythroblastosis fetalis (EP) who received intrauterine transfusions (IUT), 7 with EF or anemia without IUT, and 9 normal infants. Adult erythrocytes retained their characteristic oxygen-binding properties in the fetus (56±21 days), suggesting that a higher fetal than maternal-blood affinity for oxygen is not a prerequisite for intrauterine existence (IUT fetuses had normal growth indexes and normal acid-base values at birth). IUT infants (2-4 IUTs) had a mean P_{50} (half-saturation oxygen tension at pH 7.40) of 27.1 mm of mercury; EF infants without IUT and normals had a P_{50} of 20.8 mm of mercury, differing significantly ($p<0.0005$) from the IUT infants. A significant ($r=0.96$, $p<0.0001$) inverse relation between P_{50} and the percentage of fetal hemoglobin was noted, higher levels of fetal hemoglobin being associated with a higher oxygen affinity. Increased concentrations of red-cell 2,3-diphosphoglycerate (2,3-DPG) were found in the IUT infants. Added to the lack of correlation between oxygen-carrying capacity of cord blood and P_{50} in controls, and the significant correlation in IUT infants, this implies that, unlike the adult, the fetus does not respond to hypoxia or anemia by an increased concentration of red-cell intracellular 2,3-DPG and a shift in the oxygen-dissociation curve to the right. (47 refs.) - *K. L. Woodworth.*

Oregon Regional Primate Research Center
Beaverton, Oregon 97005

2600 **CHALMERS, THOMAS C.; & ALTER, HARVEY J.** Management of the asymptomatic carrier of the hepatitis-associated (Australia) antigen. *New England Journal of Medicine*, 285(11): 613-617, 1971.

Nearly 12,500 asymptomatic carriers of the hepatitis-associated antigen (HAA) may be found in the next year through routine testing of blood donors for HAA. Those with potentially treatable liver disease need to be differentiated from those

needing only reassurance and followup. Determination of serum transaminase aids in separating HAA-positive patients into those with acute or chronic hepatitis and those without clinically important liver disease (the latter will have normal transaminase levels). If transaminase abnormalities persist beyond 6 months and return to normality has not begun, a liver biopsy is indicated to differentiate benign persistent or unresolved hepatitis from the more serious chronic active aggressive hepatitis. Steroid therapy has been shown to have an early life-saving effect in the aggressive form of the disease. Epidemiologic data suggest that non-parenteral transmission of HAA can occur (fecal-oral, perhaps), and that very small doses can cause disease after parenteral transmission. Identified carriers should be warned of precautions to take to prevent infection of others, but restrictions are minimal. Health care workers should undergo routine screening for HAA; carriers should be especially careful (meticulous handwashing, wearing of gloves when necessary), but there is no evidence yet that they should change their careers. (49 refs.) - *K. L. Woodworth.*

National Institutes of Health Clinical Center
Bethesda, Maryland 20014

2601 **POLAND, RONALD L.; & *ODELL, GERALD B.** Physiologic jaundice: The enterohepatic circulation of bilirubin. *New England Journal of Medicine*, 284(1): 1-6, 1971.

The study evaluated the role of enterohepatic circulation of bilirubin in physiologic hyperbilirubinemia by giving to 9 full-term infants delivered by cesarean section a formula supplemented with agar (250 mg agar powder, U.S.P., in first 5 ml of each formula, beginning at 20 hours of age, for 24 doses at 4-hour intervals). Agar stabilizes bilirubin in aqueous solution, protecting it from bacterial alteration. Ten control infants received the same formula without agar. Measured daily were serum bilirubin, bilirubin excreted in stools, and weight. In contrast to the controls, serum bilirubin concentrations did not rise in the agar-fed Ss. By the sixth day, total amounts of bilirubin excreted by both groups did not statistically differ (averaged 4.0 mg/kg/day); however, agar-fed Ss excreted much more of the total bilirubin eliminated in the first 3 days, whereas controls had a progressive increase in the excretion. The simultaneous elimination of further hyperbilirubinemia and the greater excretion of bilirubin in the meconium stools suggest that agar feeding reduces hyperbilirubinemia by blocking the enterohepatic circulation of bilirubin, and that reabsorption of bilirubin from the intestine may be primarily responsible for neonatal physiologic jaundice. (33 refs.) - *K. L. Woodworth.*

*Johns Hopkins Hospital
Baltimore, Maryland 21205

2602 **GOLDBY, STEPHEN.** Experiments at the Willowbrook State School. *Lancet*, 1(7702): 749, 1971. (Letter)

Three editorials in *The Lancet* have referred to the work of Krugman et al. at the Willowbrook State School (in which MR children were infected with hepatitis in an institution where the disease was already endemic), but no editorial position was taken on the ethics of this experimental work. Krugman's study is unjustifiable, whatever the aims, because it is indefensible to give potentially dangerous infected material to children, particularly the MR, when no benefit can result to those individual children. The question of parental consent is irrelevant. The fact that Krugman and Giles state that their work conforms to the World Medical Association Draft Code of Ethics on Human Experimentation serves only to show that there is a loophole in the Draft Code, which puts the onus of consent for experimentation on children on the parent or guardian. References to this work should stop or be heavily qualified. (The editor replies that *The Lancet* accepts these criticisms. Even the experiments' hope of someday preventing hepatitis at Willowbrook and other institutions does not justify the giving of infected material to children who would not directly benefit.) (9 refs.) - *K. L. Woodworth.*

The Radcliffe Infirmary
Oxford OX2 6HE, England

2603 **CATALANO, LOUIS W., JR.; *SAFLEY, GARY H.; MUSELES, MELVIN; & JARZYNSKI, DONALD J.** Disseminated herpesvirus infection in a newborn infant. *Journal of Pediatrics*, 79(3): 393-400, 1971.

A 6-day-old Caucasian female full-term infant was admitted to the hospital because of fever and poor feeding, and disseminated Herpesvirus hominis (HVH) infection (type 1' strain) was diagnosed. A week prior to delivery the mother had herpetic vulvovaginitis, although she was free of visible lesions 24-48 hours before delivery. In the infant, high titers of type 1 HVH were isolated from throat, whole blood, and skin vesicle aspirate specimen, and from all organs obtained at necropsy. An elevated IgM level (32 mg per 100 ml) was found. No detectable HVH neutralizing or complement-fixing antibodies were present at 9 days of age. Disseminated intravascular coagulation developed 3 hours prior to death at 11 days. Interferon was present in the serum and urine at 9 days, in the urine at 10 days, and in all organs at necropsy. Interferon titers of the necropsy specimens approximately paralleled the quantities of virus present in the respective organs. The incubation period in the infant and the fact that she was symptom-free until 5 days old suggest that infection occurred during delivery. Herpesvirus was therefore probably present in the mother's birth canal despite the absence of lesions. (29 refs.) - *K. L. Woodworth.*

*Neurological Institute
New York, New York 10032

2604 **HAYES, KATHLEEN; & GIBAS, HALINA.** Placental cytomegalovirus infection without fetal involvement following primary infection in pregnancy. *Journal of Pediatrics,* 79(3): 401-405, 1971.

Reported is the first case in which the fetus was spared in the presence of placental cytomegalovirus (CMV) infection. Primary maternal infection with CMV occurred between 14 and 19 weeks of pregnancy, demonstrated by the development of CMV complement-fixing antibodies during this time, and of viruria at 25 weeks. A cervical swab at 38 weeks also yielded CMV. Development of neutralizing antibodies in the mother's serum confirmed that the infection was a primary one. A healthy male infant, weighing 2,890 gm, was delivered at 41 weeks. Culture of the placenta yielded CMV. However, CMV was not isolated from the baby despite culture of 11 urine specimens and 9 oropharyngeal swabs during the neonatal period and through the first 12 months of life. Normal values of IgM in cord serum and the absence of IgA are added evidence that the fetus was not infected. The child's development has been normal. (12 refs.) - *K. L. Woodworth.*

Royal Children's Hospital Research Foundation
Parkville, Victoria, 3052, Australia

2605 **RUDOLPH, MARTIN F.** Screening for bacteriuria in the newborn nursery: Collection of the suitable urine specimen. *Journal of Pediatrics,* 79(3): 463-466, 1971.

A 2-chambered urine collector (U-Bag) was used in screening 110 female and 90 male full-term healthy newborns for bacteriuria in a community hospital. The adhesive plastic urine collector was designed to minimize exposure of the urine to contamination from the perineum and genitals. A laboratory technician daily applied the bag to the grossly clean perineum of each infant. The collector bag was removed after 1 hour if the child failed to void. Quantitative bacterial counts were performed on all specimens using the streak-plate method. Specimens suitable for analysis were obtained from 81% of the infants. In only 6% of the specimens did bacterial counts exceed 10^5 organisms per ml, and in no instance did this degree of bacteriuria persist in 3 consecutive specimens. (5 refs.) - *K. L. Woodworth.*

70 Deer Hill Avenue
Danbury, Connecticut 06810

2606 **GATTI, RICHARD A.; & GOOD, ROBERT A.** Follow-up of correction of severe dual system immunodeficiency with bone marrow transplantation. *Journal of Pediatrics,* 79(3): 475-479, 1971.

Immunologic responses and lymphoid systems were reconstituted in a child with sex-linked severe combined immunodeficiency. The 6-month-old infant with lymphopenic hypogammaglobulinemia received the initial bone marrow transplantation on August 24, 1968; 8 weeks later a second transplant from the same donor was required to correct an aregenerative pancytopenia consequent to the initial transplant. Since that time the immunologic and hematologic

reconstitution has remained intact, and the S is clinically healthy. A recent EB virus infection caused only mild symptoms, which subsided spontaneously. A new self-regenerating marrow was established in the recipient. His persistent chimeric state is reflected in the female karyotype patterns of the marrow cells and peripheral lymphocytes, a change from blood group A(S's original group) to O(blood group of the donor), and the presence of a hybrid $A_1 Le^b$ molecule on the surface of the O erythrocytes. (7 refs.) - *K. L. Woodworth.*

University of Minnesota
Variety Club Heart Hospital
Minneapolis, Minnesota

2607 **PELC, S.** E.E.G. in herpes-simplex encephalitis. *Lancet,* 2(7719): 320, 1971. (Letter)

EEG studies in a proven case of herpes-simplex encephalitis corroborate previously reported observations elsewhere that patients with herpes-simplex encephalitis develop periodic complexes in the EEG between the second and fifteenth days of the disease. In the case cited, the EEG showed right-temporal slow waves on days 5, 8, and 10, but periodic patterns on days 13 and 16. The periodic complexes, at 2-3 second intervals, were mostly confined to the right side of the brain. Unilateral EEG periodicity or periodicity preponderantly on one side may be the earliest distinctive EEG features of herpes-simplex encephalitis, since others have also reported unilateral periodicity in at least 3 cases. (5 refs.) - *K. L. Woodworth.*

Universite Libre de Bruxelles
Belgium

2608 **SCHIFF, GILBERT M.; & RAUH, JOSEPH L.** Rubella control. *American Journal of Diseases of Children,* 122(2): 112-116, 1971.

Tools presently exist for effective control and eventual eradication of the rubella problem. A rubella control program should include establishment of adequate facilities to provide for viral and serological tests needed for laboratory diagnosis and immune status determination, proper management of suspected cases of acquired and congenital rubella, routine determination of rubella immune status in females of childbearing age, intelligent use of rubella vaccines, and proper management of exposed, pregnant women. Intelligent use of rubella vaccines consists in immediate mass vaccination of children aged 1 through 10 by school systems, clinics, and private physicians, adoption of rubella vaccination as part of routine pediatric care, selected vaccination of susceptible postmenarchial females who are not pregnant and will avoid pregnancy for at least 8 weeks following vaccination, and postpartum vaccination of susceptible pregnant women. (26 refs.) - *B. J. Grylack.*

Christ Hospital Institute of Medical Research
Cincinnati, Ohio 45219

2609 **NAHMIAS, ANDRE J.** Herpes genitalis: Treatment, effect on pregnancy. *Journal of the American Medical Association,* 217(9): 1250, 1971.

There is at present no adequate therapy in the United States for genital herpes simplex infection. Idoxuridine (Stoxil), smallpox vaccination, and herpes nongenital type 1 virus vaccine have been generally unsuccessful. A type 2 genital virus vaccine available in West Germany is more effective, but it will not be used in the United States until the association of type 2 herpes with cervical cancer is clarified. The woman with genital herpes should have frequent Papanicolaou examinations for cervical cancer detection. An increased risk of abortions and congenital infections exists in the woman with genital herpes. If pregnancy occurs, an effort to detect evidence of herpetic infection in the last few weeks of pregnancy should be made, because the risk of congenital herpes is greater if the disease is active at the time of delivery. (No refs.) - *B. J. Grylack.*

Emory University School of Medicine
Atlanta, Georgia

2610 **SOUTH, DALE R., JR.; & NAHMIAS, ANDRE J.** Herpes genitalis: Risk to newborn. *Journal of the American Medical Association,* 217(9): 1250, 1971.

A 24-year-old woman, gravida III, para 1, who had a genital herpes simplex infection acquired

from her husband, requested consultation on this problem 2 months prior to the expected date of delivery of her child. The patient was closely observed during these 2 months, as advised, and the pathologist was unable to identify evidence of a herpes infection in a Papanicolaou smear. An elective cesarean section was performed after 2 months because the parents were concerned about the possibility of infection in the child and wanted a tubal ligation performed. Moreoever, a questionable flare-up of the infection had occurred approximately 2 weeks prior to delivery. The baby has shown no evidence of congenital disease, but the parents continue to have recurrent genital herpetic lesions. (No refs.) - *B. J. Grylack.*

2611 **HORSTMANN, DOROTHY M.** What risk in pregnancy diagnosed three months after rubella vaccination? *Journal of the American Medical Association,* 217(9): 1250-1251, 1971.

A case was reported of a woman who was given rubella vaccine 3 months before the diagnosis of pregnancy was made, and concern was voiced in regard to possible damage to the fetus. It would be useful to know whether the woman had a serologic test to determine her susceptibility to rubella and the relationship between dates of conception and vaccination. If conception took place 2 or 3 months before vaccination, there should be no concern about the fetus. In a susceptible person the period of virus shedding from the throat (which reflects active infection) is primarily between postvaccinal days 10 and 20, although it occasionally lasts as long as 28 to 30 days. This type of problem should emphasize the need for serological testing, preferably the standardized hemagglutination inhibition (HAI) test, before rubella vaccine is administered to a woman of childbearing age. (No refs.) - *B. J. Grylack.*

Yale University School of Medicine
New Haven, Connecticut

2612 **BHOYROO, S. K.; & STORRS, C. N.** Adult and fetal haemoglobin. Haemolytic disease of the newborn. *Archives of Disease in Childhood,* 46(248): 570-571, 1971.

A series of 14 rhesus affected babies who had not received intraperitoneal transfusion was studied to determine any disproportionate increase in adult hemoglobin. According to estimates made by the alkaline denaturation method of Singer and by the Kleinhauer technique on cord blood, no difference in the proportions of fetal and adult hemoglobin was found as compared to cord blood of normal infants of similar gestational age. The results indicate that the Kleihauer technique can be used with accuracy to establish the hemoglobin proportions in cord blood. Given the fact that there is a wide range in normal values for both rhesus affected babies and normal controls, these proportions may be useful as a measure of success of intraperitoneal transfusion given before birth. (9 refs.) - *B. J. Grylack.*

Institute of Child Health, Hammersmith Hospital
London W. 12, England

2613 Rh problem in therapeutic abortion. *Medical World News,* 12(17): 36G, 1971.

Unless the father is Rh negative or the mother has already been immunized, every Rh negative woman who had an induced abortion should receive Rh immune globulin. Significant feto-maternal hemorrhages were found in 29 of 404 women who had suction-curretage abortions between the seventh and sixteenth weeks of gestation. Of 202 women who had saline abortions, 41 had significant hemorrhaging, and 25 of these had significant feto-maternal hemorrhage after the amniocentesis and prior to the injection of hypotonic saline. Incidence and volume of hemorrhaging increased with the length of pregnancy and was greater following saline abortion. Of the 1 million abortions which will be performed annually by 1972, approximately 150,000 will be performed on Rh negative women. Chances of becoming sensitized for these women are 14 to 17% in the case of a full-term Rh positive pregnancy and 3 to 4% in the case of spontaneous abortion followed by curretage. (No refs.) - *J. K. Wyatt.*

2614 **O'TOOLE, RICHARD D.; THORNTON, GEORGE F.; MUKHERJEE, MRINAL K.; & NEOGY, KRIPA N.** Cerebrospinal fluid immunoglobulins in bacterial meningitis. *Archives of Neurology,* 25(3): 218-224, 1971.

The potential contribution of anticapsular antibody to host defenses in pneumococcal meningitis is evaluated. Ss were 31 patients with pneumococcal, meningococcal and tuberculous meningitis from whom samples of serum and cerebrospinal fluid (CSF) were taken. Pneumococci were isolated from the CSF of every patient and were characterized by colonial morphology, bile solubility, sensitivity to ethylhydrocuprein and serologic typing. Each patient's serum was tested within 3 weeks of hospital admission against the pneumococcus cultured from his CSF. The IgM, IgG and IgA were quantified by a radial diffusion method using antiserum agar plates and antigen standards. Pneumococcal C substance, Type I pneumococcal polysaccharide and rabbit antiserum to Type I were made available. The presence or absence of antibody to Type I pneumococcal capsular polysaccharide in CSF was determined using a radioimmunoassay. Fifteen of 31 patients with pneumococcal meningitis were cured, 14 died and 2 showed residual neurologic deficits. Patients with antibody had fewer CSF leukocytes and significantly lower CSF pressures. Ss differed so strikingly with respect to possession (or lack) of antibody and cure that it is suggested antibody and cure may be causally related. (35 refs.) - *T. C. Turner.*

Seattle Veterans Administration Hospital
Seattle, Washington 98108

2615 **KLIMAN, ALLAN; REID, NANA R.; LILLY, CONSTANCE; & MORRISON, JEAN.** Hepatitis-associated antigen (Australia antigen) in Massachusetts blood donors. *New England Journal of Medicine*, 285(14): 783-785, 1971.

The results of large-scale hepatitis-associated antigen (HAA) testing of Massachusetts blood donors are reported. Since August 1970 all bloods collected by the Massachusetts Red Cross Blood Program were tested for HAA using Austect reagents. From August to October agar gel diffusion technic only was used. From October to March, counter immunoelectrophoresis (CIEP) was used. The pilot tube sample for HAA testing was either serum or ethylenediamine tetra-acetate (EDTA) plasma and all tests were performed within 48 hours of blood collection. Blood donors who were HAA positive were recalled for further testing. Serum glutamic oxalacetic transaminase (SGOT), serum glutamic pyruvic transaminase (SGPT) and bilirubin were also tested on each retest sample. Of 106,294 consecutive blood donors, 114 were positive. The number of donors found to be positive in each month was constant. Approximately one-third of these donors had other liver-function abnormality. In 75 HAA-positive donors, the SGOT was above 50 Karmen U in 22, SGPT above 35 International U in 6 and bilirubin above 1.0 mg per 100 ml in 5. Thirty of the 114 donors were prisoners in state or county correctional institutions; there were only 8 females among the 114. Finding HAA-positive donors represents a new form of public health screening. HAA-positive donors differ from the general population and in this finding is the hope that the overall risk of hepatitis from blood collection will be diminished as high-risk populations are identified. There is a predominance of males among HAA-positive donors. (8 refs.) - *T. C. Turner.*

812 Huntington Avenue
Boston, Massachusetts 02115

2616 **PASHAYAN, H.; PRUZANSKY, D.; & PRUZANSKY, S.** Are anticonvulsants teratogenic? *Lancet*, 2(7726): 702-703, 1971. (Letter)

In addition to 32 children previously reported with cleft lip and palate, with or without other malformations, born to epileptic mothers on anticonvulsant drugs, 4 more cases have been described. Although the argument against anticonvulsants is weakened by the history of facial clefts in 1 of these families and by the birth of normal siblings to a mother who was taking anticonvulsants, the suspicion remains that these drugs are a contributory factor in the etiopathogenesis of clefts. In view of the finding that folic-acid deficiency has caused cleft palate in rats and mice and that anticonvulsants are known to act as folic-acid antagonists, the combination of vitamin supplements and anticonvulsants merits examination in laboratory animals. (10 refs.) - *B. J. Grylack.*

Center for Craniofacial Anomalies
P.O. Box 6998
Chicago, Illinois 60680

2617 **SCHULKIND, MARTIN L.; ALTEMEIER, WILLIAM A., III; & AYOUB, ELIA M.** A comparison of ampicillin and

chloramphenicol therapy in *Hemophilus influenzae* meningitis. *Pediatrics*, 48(3): 411-416, 1971.

A comparison was made of 16 children receiving chloramphenicol and 21 receiving ampicillin, all of whom were being treated for *Hemophilus influenzae* meningitis. The parameters used to evaluate the response to therapy included duration of fever, changes in cerebrospinal fluid (CSF) laboratory values, incidence of residual neurological sequelae, and mortality rates. Those treated with ampicillin tended to have fever longer than those receiving chloramphenicol, with a difference significant at a p value of .002. There was no evidence that the variation in antibiotic regimens influenced the return of CSF laboratory values to normal, the incidence of subdural effusions, or the incidence of neurological sequelae. Two patients who had been given each drug did not survive the illness. These observations indicated that a prolonged febrile course may be expected during effective treatment with ampicillin, but there is no indication for replacement of ampicillin therapy as long as other criteria show evidence of a favorable response to therapy. (19 refs.) - *B. J. Grylack.*

University of Florida College of Medicine
Gainesville, Florida 32601

2618 U. S. Public Health Service. Medical aspects of childhood lead poisioning. *Pediatrics*, 48(3): 464-468, 1971.

The U.S. Public Health Service has recommended that screening programs for the prevention and treatment of lead poisoning in children include all those who are between the ages of 1 and 6 years, who live in old, poorly maintained houses, or who are exposed to other special local conditions involving lead hazards. Blood lead concentrations of 40 μg or more per 100 ml of whole blood, determined on 2 separate occasions, should be considered evidence suggestive of undue absorption of lead, either past or present. Children with greater than 80 μg lead per 100 ml of whole blood should be considered unequivocal cases of lead poisoning and should be treated as medical emergencies. Children with lead values of 50 to 79 μg per 100 ml of whole blood should be referred immediately for evaluation as possible cases of lead poisoning. This recommendation applies as well to children with lead values between 40 and 49 μg per 100 ml. All children who are diagnosed with lead poisoning should receive immediate treatment, must be removed from the source of lead exposure and followed carefully until at least 6 years of age, and should be given adequate neurological and psychological assessment and additional clinical and laboratory evaluation. (16 refs.) - *B. J. Grylack.*

U.S. Public Health Service
Cincinnati, Ohio 45202

2619 **YEUNG, C. Y.; TAM, L. S.; CHAN, ANGELINE; & LEE, K. H.** Phenobarbitone prophylaxis for neonatal hyperbilirubinemia. *Pediatrics*, 48(3): 372-376, 1971.

One hundred and thirty-three term infants born of group O mothers were included in a study designed to test the efficacy of phenobarbitone as a prophylaxis in reducing the incidence and severity of neonatal jaundice in newborns. Of the 3 study groups which were created, 1 consisted of 45 control infants receiving no medication, another of 44 infants born of mothers who received 30 mg of the drug every night for an average of 2 weeks prior to delivery, and a third of 44 infants receiving 5 mg of phenobarbitone every 8 hours for an average of 3.5 days beginning 6 to 8 hours after birth. On the basis of 6 days' measurement, infants receiving prophylactic phenobarbitone were found to have a markedly lower incidence of hyperbilirubinemia as compared to the control Ss, with an observed difference reaching statistical significance ($x^2 = 13{\cdot}29$, $p = <0.01$) These treated infants had a lower incidence of hyperbilirubinemia than those born of treated mothers, although this difference did not attain a significant probability limit ($X^2 = 8.05$, $p = <.10$). Enhancement of liver functions probably forms the basis of the efficacy of phenobarbitone in lowering the serum bilirubin in newborn infants. (23 refs.) - *B. J. Grylack.*

Paediatric 'B' Unit
Queen Elizabeth Hospital
Kowloon, Hong Kong

2620 **CHISOLM, J. JULIAN, Jr.** Increased lead absorption: Toxicological considerations. *Pediatrics*, 48(3): 349-352, 1971.

A working knowledge of the pertinent time-dose-response relationships of lead absorption is necessary to deal with its biologic effects. A recent statement by the U.S. Public Health Service has placed great emphasis on blood lead levels. While the relationship between blood lead levels and current exposure has not been defined precisely, exponential increases in "dose" appear to be reflected by arithmetic increases in blood lead concentration. The recommendations made by the Public Health Service for pediatric care vary according to the estimate of current risk provided by blood lead levels. Levels less than 40 μg lead are assumed to indicate a negligible risk, with normal daily dietary intake probably accounting for most of the intake. Levels greater than 40 μg signify some additional source of intake. Levels from 50 to 79 μg indicate an accelerating risk, an increasing probability of demonstrable metabolic impairment in heme synthesis, and the possibility of ill-defined symptoms of illness. Levels above 80 μg appear to present an unacceptable risk, particularly if long sustained. The most serious limitation imposed upon good medical management in children as well as adults is the very limited availability to the general public of reliable lead analyses in blood and urine. (23 refs.) - *B. J. Grylack.*

Johns Hopkins University School of Medicine
Baltimore, Maryland

2621 **LIN-FU, JANE S.** Diagnostic and screening procedures for lead poisoning. *Pediatrics,* 48(3): 488-489, 1971. (Letter)

In response to criticism by the American Academy of Pediatrics Committee on Environmental Hazards regarding the use of 24-hour urine studies as mentioned in the U.S. Public Health Service report on lead poisoning, it has been pointed out that "screening" and "diagnosis" are not synonymous procedures. Blood lead determinations should be used as screening tests, while 7 tests including 3 requiring 24-hour urine collection were recommended for diagnosis in the case of children with lead levels of 50 to 79 μg/100 ml of whole blood. The Committee also argued that a blood lead concentration of over 40 μ/100 ml does not necessarily mean that the child is poisoned. The Public Health Service report did not state or imply that correlation but reasoned that a lead level of 40 μg/100 ml of whole blood determined on 2 occasions represented an undue absorption of lead. It is important to investigate these children so that exposure can be terminated before poisoning does occur. (No refs.) - *B. J. Grylack.*

Maternal and Child Health Service
Rockville, Maryland 20852

2622 **LOURIE, REGINALD S.; & WEHRLE, PAUL F.** Prevention of lead paint—or prevention of pica? *Pediatrics,* 48(3): 490-491, 1971. (Letter)

The American Academy of Pediatrics Committee on Environmental Hazards has been criticized for failing to bring attention to the causes of pica in children in its report on lead poisoning. In the majority of cases, pica is probably related to unavailable mothering. The pediatrician usually has to turn to the social services to help to define the basis of the mother's unavailability and to set in motion the corrective forces available in the community. A range of resources must be available, including those which can reach into the home and deal with problems frequently imposed by poverty. This approach uncovers the overwhelmed, dependent and helpless, mentally ill or MR, and alcoholic and/or pleasure-oriented teen-age mothers, all of whom can utilize assistance. Unfortunately, these extensive resources may not be available in all communities or may not be readily mobilized in time to avert the consequences of lead exposure. (3 refs.) - *B. J. Grylack.*

Children's Hospital of D.C.
Washington, D.C. 20009

2623 **BARRETT, PETER V. D.** Hyperbilirubinemia of fasting. *Journal of the American Medical Association,* 217(10): 1349-1353, 1971.

Five normal Ss and 5 Ss with hepatic dysfunction were fasted for 44 to 48 hours in order to determine the effect of fasting on serum bilirubin concentration. One abnormal S had Gilbert's syndrome and an atypical form of hereditary spherocytosis, 2 had primary biliary cirrhosis, 1 had probable alcoholic cirrhosis, and 1 had Gilbert's syndrome alone. At the end of the

fasting period, all 10 Ss showed an increase in total serum bilirubin (TSB) and direct bilirubin (DB) concentrations. There was no significant difference between the normal S and the patients with hepatic dysfunction in regard to the percent increase of the TSB ($p<0.05$) or the DB ($p<0.15$). A diurnal rhythm of the TSB was demonstrated in a patient with primary biliary cirrhosis, but it was suppressed by nocturnal feedings. Hemolysis and bilirubin retention as a result of diminished renal excretion were excluded as significant causes of hyperbilirubinemia. Fasting hyperbilirubinemia is clinically significant especially in conditions such as hyperemesis gravidarum or congenital pyloric stenosis, which are often accompanied by jaundice. (16 refs.) - *B. J. Grylack.*

1000 W. Carson Street
Torrance, California 90509

2624 **STENCHEVER, MORTON A.; & PARADA, MALCOLM.** Management of the severely affected Rh-isoimmunized patient. *American Journal of Obstetrics and Gynecology*, 111(2): 274-279, 1971.

Experience with 235 severely Rh-isoimmunized patients over a 6-year period has been summarized, with particular attention paid to a comparison of results obtained in the transfused and nontransfused cases. Of the 235 sensitized patients, 182 were found to have a difference in optical density (δO.D.) at 450 μm of never more than 0.14 and were not considered for any treatment except early delivery in appropriate cases. Of the 53 cases with an observed δO.D. of 0.15 and above, 29 were given 41 intrauterine transfusions, resulting in the delivery of 9 healthy infants, 7 neonatal deaths, and 13 stillbirths. Twenty-four patients with δO.D. values of 0.15 or above were not treated with intrauterine transfusion, resulting in 14 infants who survived, 6 stillbirths, and 4 neonatal deaths. Transfusion should be given to fetuses with a δO.D. of greater than 0.25 who are not salvageable by delivery and intensive postnatal care at the time of evaluation. An attempt should be made to continue these babies *in utero* until 35 to 36 weeks' gestation whenever possible. (8 refs.) - *B. J. Grylack.*

50 North Medical Drive
Salt Lake City, Utah 84112

2625 **ALTSHULER, GEOFFREY; & McADAMS, A. JAMES.** Cytomegalic inclusion disease of a nineteen-week fetus. Case report including a study of the placenta. *American Journal of Obstetrics and Gynecology*, 111(2): 295-298, 1971.

A unique case of cytomegalic inclusion disease was documented in a 19-week fetus. The male infant died 1 hour after delivery. Routine histologic examination of the placenta and fetus indicated the presence of cytomegalovirus. The mother was found to have viral hepatitis; however, no cytomegalic inclusions were demonstrated on liver biopsy. The placental lesion in this case was characterized by a focal villitis and the presence of plasma cells in the lesion, in addition to the cytomegalic virus itself. The cause of premature delivery was not clear. This is the only case known to be reported of cytomegalovirus infection of a fetus of this degree of immaturity. (8 refs.) - *B. J. Grylack.*

General Hospital
Cincinnati, Ohio

2626 **KIMBALL, ANNE C.; KEAN, B. H.; & FUCHS, FRITZ.** Congenital toxoplasmosis: A prospective study of 4,048 obstetric patients. *American Journal of Obstetrics and Gynecology*, 111(2): 211-218, 1971.

The frequency of congenital toxoplasmosis was determined in 4,048 infants delivered at 1 hospital center over an 18-month period. The dye test (DT) was negative (less than 1/16) on 2,765 patients and positive on 1,283. The complement fixation test (CFT) was positive on 369. During pregnancy, 6 obstetric patients converted from DT negative to positive, and 2 of the 6 transmitted toxoplasmosis to their infants. Of the 1,283 patients who had positive tests for toxoplasma antibodies on prenatal blood specimens, 17 showed a substantial rise in titer during pregnancy, and 1 transmitted toxoplasmosis to her infant. The fact that 68% of the patients lacking the protection of toxoplasma antibodies were at risk was consistent with previous studies. All pregnant women should be advised to eat only well-cooked meat and to avoid cats and cat feces. Routine quantitative immunoglobulin M determination on all cord bloods is recommended for its value in providing early indication of

infection with toxoplasma gondii as well as in detecting infants with a wide spectrum of intrauterine infections. (14 refs.) - *B. J. Grylack.*

The New York Hospital-Cornell Medical Center
New York, New York

2627 **EARNEST, M. P.; GOOLISHIAN, H. A.; CALVERLEY, J. R.; HAYES, R. O.; & HILL, H. R.** Neurologic, intellectual, and psychological sequelae following western encephalitis. *Neurology*, 21(9): 969-974, 1971.

Observations on the sequelae of western encephalitis (WE) are extended to 35 Ss in Hale County, Texas; attempts are made to resolve disparity in findings of previous studies and to add to our knowledge of post encephalitic sequelae, an estimate of the economic costs of brain damage among infants and children. Each S performed tests of balance and co-ordination and was given a physical examination. One year later 22 Ss were relocated and given the WAIS, WISC, Wechsler Preschool and Primary Scale of Intelligence (WPPSI), or Stanford-Binet test, Goodenough Draw-A-Person Test, Bender-Gestalt visual-motor test and Raven progressive matrices. Information on personality, social adjustment and possible psychomotor difficulties was also obtained. A closely matched control group was obtained and tested. From the 35 Ss, 12 had significant sequelae. Seven were MR, had recurrent convulsions, unilateral/bilateral spastic weakness; minimal brain dysfunction was found in 4; and 1 had severe speech and hearing deficit. Incidence and severity of sequelae were closely related to age at onset of WE. The costs for each epidemic of WE from 1963-1966 were $9000. Total economic costs of lost productivity and institutional care following WE are estimated at $320,000. Special intensive effort should be made to prevent sequelae, such as education of mothers to protect infants from mosquitoes, provide mosquito netting for cribs and aerosol cans of insecticide to use during high vector mosquito activity. (26 refs.) - *T. C. Turner.*

University of Pennsylvania
Department of Neurology
Philadelphia, Pennsylvania 19104

2628 **HUISMAN, T.H.J.; SCHROEDER, W.A.; CHARACHE, S.; BETHLENFALVAY, NICHOLAS, C.; BOUVER, N.; SHELTON, J. ROGER; SHELTON, JOAN BALOG; & APELL, GERALD.** Hereditary persistence of fetal hemoglobin. *New England Journal of Medicine*, 285(13): 711-716, 1971.

In 30 members of 3 Negro families, determination of Gγ to Aγ chains ratio of human fetal hemoglobin agreed with results from other Ss of the same types. Four groups of conditions were analyzed: (1)heterozygotes; (2)homozygotes for the hereditary persistence of fetal hemoglobin; (3)β-thalassemia trait; and (4)β-thalassemia trait plus the hereditary persistence of fetal hemoglobin. An unknown factor seems to change the expression of the γgenes in homozygotes for the hereditary persistence of fetal hemoglobin compared to that of the ordinary heterozygotes. In group 4 individuals, augmented synthesis of fetal hemoglobin may be guided by the β-thalassemia gene. (19 refs.) - *B. Berman.*

Medical College of Georgia
Augusta, Georgia

2629 **BUNN, H. FRANKLIN.** Hemoglobin heterogeneity. *New England Journal of Medicine*, 285(13): 746-747, 1971.

That structural differences occur between comparable adult and fetal proteins is shown in the transition from hemoglobin F to hemoglobin A. Elevated levels of hemoglobin F have occurred as incidental findings in otherwise normal families. Hereditary persistence of fetal hemoglobin is apparently heterogeneous. Various findings indicate the presence of multiple γ-chain genes and suggest that the alanine-glycine heterogeneity results from 2 or more separate genes rather than from ambiguous translation of the messenger. α-chain synthesis may also be governed by more than one gene. These considerations are important in a number of acquired and inherited disorders, among them α- and β-thalassemia. (4 refs.) - *B. Berman.*

No address

2630 **STOKES, JOSEPH, JR.; WEIBEL, ROBERT E.; VILLAREJOS, VICTOR, M.; ARGUEDAS, JORGE A.; BUYNAK, EUGENE B.; & HILLMAN, MAURICE R.**

Trivalent combined measles-mumps-rubella vaccine. *Journal of the American Medical Association*, 218(1): 57-61, 1971.

A combined trivalent Moraten measles, Jeryl Lynn mumps, and HPV-77 duck rubella vaccine was tested clinically in 715 children between the ages of 7 months and 7 years who were initially without antibody against any of the 3 viruses. The main goals of the study were to help to simplify administration of vaccines, to reduce costs, and to minimize the required number of contacts between patient and physician. The overall seroconversion rate for measles was 96%, and the geometric mean (GM) titer was 1:38.5. The same values for mumps and rubella were 95% and 1:6.8, and 94% and 1:27.8. respectively. There was no detectable suppression of the height of antibody titer following combined vaccine as compared with monovalent vaccines. HPV-77 duck rubella and Jeryl Lynn mumps vaccines cause remarkably little clinical reaction, while Moraten measles may cause mild febrile reactions, rash, and generalized malaise. There was no apparent alteration of the clinical reactivity by the combined administration of the 3 vaccines in a single dose. Arthritis and arthralgia, prominent features of adult rubella, were absent. The combined triple vaccine provides a simple, safe, and effective immunization procedure using a single vaccine dose against 3 important diseases in children who have not been immunized. (8 refs.) - *B. J. Grylack.*

University of Pennsylvania School of Medicine
Philadelphia, Pennsylvania

2631 **SINNIAH, D.; TAY, L. K.; & DUGDALE, A. E.** Phenobarbitone in neonatal jaundice. *Archives of Disease in Childhood*, 46(249): 712-715, 1971.

Eighty-three jaundiced newborn infants were studied in an assessment of the effectiveness of postnatal treatment of neonatal hyperbilirubinemia with phenobarbitone. Of 60 normal weight infants, 28 received phenobarbitone and 32 acted as controls. At 48 hours after treatment with phenobarbitone, the mean serum bilirubin level of the treated infants was significantly lower than that of controls. Exchange transfusions were required in 6 of the 32 controls but not in any of the treated cases. There were no significant differences in the mean serum bilirubin levels or in the exchange transfusion rates between 13 treated and 10 control low-birthweight infants. No increase in mortality rate was found among infants treated with phenobarbitone, and no serious adverse effects were noted apart from drowsiness and difficulty in feeding, which occasionally persisted for as long as 2 days after treatment was discontinued. (14 refs.) - *B. J. Grylack.*

University of Malaya
Kuala Lumpur, Malaysia

2632 **SAVILAHTI, E.; PELKONEN, P.; & VISAKORPI, J. K.** IgA deficiency in children. A clinical study with special reference to intestinal findings. *Archives of Disease in Childhood*, 46(249): 665-670, 1971.

Twenty-six children with immunoglobulin (Ig) A deficiency were studied, with special attention directed towards intestinal findings. Autoimmune disease, malabsorption syndrome, and recurrent respiratory infections were 3 separate disease entities found among the 26 children. In 21 cases, no IgA was detectable; trace amounts were seen in 5 cases. In the majority of patients, the intestinal absorptive function was normal as studied by fecal fat and D-xylose excretion tests. Steatorrhea was evidenced in 2 patients, 1 of whom also had low urinary D-xylose excretion. In both patients with steatorrhea and in 1 other patient, small intestinal biopsy showed total villous atrophy. Disaccharidase activities were measured from 8 biopsy specimens with normal villous structure, and selective lactase deficiency was noted in 2 of these; neither patient developed symptoms after ingestion of lactose, however. Since this sample was a highly selective group of hospital patients, no definite conclusions were made regarding the prevalence of various diseases in IgA-deficient children. (34 refs.) - *B. J. Grylack.*

Children's Hospital, University of Helsinki
Helsinki, Finland

2633 **TURNER, G. C.; FIELD, ANNE M.; LASHEEN, RAGAA M.; TODD, R. M.; WHITE, G.B.B.; & PORTER, A. A.** SH (Australia) antigen in early life. *Archives*

of Disease in Childhood, 46(249): 616-622, 1971.

A case involving the transmission of serum hepatitis (SH) antigen from a mother to her infant has been reported. The baby, born during the antigenemic phase of maternal hepatitis, developed positive tests for antigen and raised serum alanine aminotransferase values after an interval corresponding to the incubation period of serum hepatitis. When the child was almost 2 years old, the test for antigen was still positive at a dilution of 1/64 by complement fixation, but the immunoglobulin M level had declined from an abnormally high level to normal limits, and the alanine transferase value (125 units/100 ml) was at the lowest level encountered since an abnormality was first detected. Throughout the investigation, the child remained well clinically. In contrast to the usual findings in an adult patient, the serological and electron microscope findings in this child suggested such a slow development of the immunological response that it was still ineffective in the elimination of antigen at age 2. The importance of antigen-associated infection in early life lies not only in its causal relation to liver disease but also in its possible significance in the genesis of long-continued carriage of SH virus. (22 refs.) - *B. J. Grylack.*

Public Health Laboratory
Liverpool L3 5SU, England

2634 **DUDGEON, J. A.** Cytomegalovirus infection. *Archives of Disease in Childhood,* 46(249): 581-583, 1971.

In 1957, cytomegalovirus (CMV) was first isolated in human tissue from patients with clinical illnesses resembling what is now know as cytomegalic inclusion disease (CID). Since that time, improved virological techniques have aided in the diagnosis and management of CID. A 2-year collaborative study recently showed that 90% of children found to be excreting virus without central nervous system disease progressed normally. The majority of cases of CMV infection presenting in the newborn and in the early weeks of life are probably congenital in origin, but it is difficult to determine at what stage in intrauterine life infection is acquired. Some infections are probably acquired quite late in pregnancy and possibly by the cervical-amniotic, as opposed to the hematogenous, route. Structural malformations of the type seen in congenital rubella, such as heart disease and cataracts, are very uncommon in CID. This factor could be used in support of the argument that fetal infection with CMV usually occurs after the period of organogenesis. Further studies are needed to ascertain knowledge of the pathogenesis of intrauterine CMV infection. (18 refs.) - *B. J. Grylack.*

Hospital for Sick Children
London W. C. 1, England

2635 **ELDER, H. A.; SANTAMARINA, B. A. G.; SMITH, S.; & KASS, E. H.** The natural history of asymptomatic bacteriuria during pregnancy: The effect of tetracycline on the clinical course and the outcome of pregnancy. *American Journal of Obstetrics And Gynecology,* 111(3): 441-462, 1971.

All 9,156 patients who registered for prenatal care at a municipal hospital over a 2.5-year period were tested for bacteriuria, and tetracycline and placebo were given to groups of bacteriuric patients in order to evaluate the effect of such treatment on the clinical course and the outcome of pregnancy. One hundred and thirty-three bacteriuric patients were treated with tetracycline and 148 with placebo. Among non-bacteriuric control Ss, 147 received tetracycline, and 132 placebos. Twenty-one percent of the patients whose bacteriuria did not clear with tetracycline or who had a recurrent episode of bacteriuria after they had their urine cleared with tetracycline and 18% of the patients who developed symptomatic pyelonephritis were delivered of premature infants, whereas only 10% of the tetracycline-treated patients who remained free of bacteriuria were delivered of premature newborns. Bacteriuric patients whose bacteriuria had cleared had a recurrence rate prior to delivery which was 8 times greater than that of non-bacteriuric controls. Symptomatic pyelonephritis occurred in 16% of bacteriuric patients and was noted only in those with preexisting bacteriuria, except for 1 case. There were no maternal complications attributable to tetracycline, and one third of the offspring had staining of the teeth but no adverse effects on growth and development, rate of dental caries, or hypoplasia of dental enamel due to tetracycline. (22 refs.) - *B. J. Grylack.*

Loma Linda University
Loma Linda, California 92354

2636 **PRINCE, A. M.; SZMUNESS, W.; MILLIAN, S. J.; & DAVID, DAVID S.** A serologic study of cytomegalovirus infections associated with blood transfusions. *New England Journal of Medicine*, 284(20): 1125-1131, 1971.

The hypothesis that cytomegalovirus infections may be transmitted by blood transfusion is supported but not proven. In 152 patients the development of complement-fixing antibody was determined. Seven per cent of 59 patients who received only a single unit of blood, 21% of 72 patients with multiple transfusions, and 52% of 21 immunosuppressed transplant recipients exhibited seroconversion an average of 8 weeks after transfusion. The immunosuppressed patients demonstrated a significantly enhanced immune response. Seroconversion risk correlated with the volume of blood transfused, but not with the use of fresh blood or with the presence of preexisting complement-fixing antibody. Post-transfusion hepatitis, mostly subclinical, developed in 23 patients, of whom SH antigen occurred in 4 and cytomegalovirus in 8. Twenty-two patients with seroconversion evidenced no hepatitis. (33 refs.) - *S. Glinsky.*

New York Blood Center
New York, New York 10021

2637 **SCANLON, JOHN.** Bilirubinemia and gut flora in infants. *New England Journal of Medicine*, 284(21): 1218, 1971. (Letter and reply)

Early colonization of the newborn infant by intestinal bacterial flora does not seem to protect the infant from idiopathic hyperbilirubinemia. Previous authors had hypothesized the absence of intestinal bacterial flora as an essential part of the newborn infant's gastrointestinal physiology. The hospital records of 6 full-term infants with bacteriologically positive gastric aspirates at birth were examined for average bilirubin levels. The average daily bilirubin values on the second, third, and fourth days were 8.85 mg, 11.8 mg and 11.4 mg/100 ml, respectively. The infants' orally ingested intestinal flora did not prevent hyperbilirubinemia. In answer the original authors suggest that bacterial action of bilirubin is a function not only of the degree of colonization but also of the species of bacteria. They concur in the suggestion that study of the bacterial flora of the gastrointestinal tract might illuminate the fate of bilirubin in the intestinal tract. (5 refs.) - *S. Glinsky.*

Boston Hospital for Women
Boston, Massachusetts

2638 **LUCAS, R. T., JR.** Rubella-measles. *Journal of the American Medical Association*, 215(7): 1156, 1971. (Letter)

Public confusion regarding measles and German measles can be corrected by means of a simple editorial expedient. In all press releases and public information the term of rubella should be followed by the term German measles in parenthesis to indicate that the latter is an inferior term. (1 ref.) - *S. Glinsky.*

No address

2639 **PLOTKIN, STANLEY A.** Rubella vaccination. *Journal of the American Medical Association*, 215(9): 1492-1493, 1971. (Editorial)

The current mass rubella vaccination program in the United States may not prevent the spread of wild rubella virus from children to their mothers. The argument of herd immunity is not demonstrable. Also, the immunologic defect in present strains seems to be the production of insufficient immunity on the nasopharyngeal mucosa, where the wild virus infects. An alternative to mass vaccination is the specific and total vaccination of prepubescent girls, which permits natural immunization by wild virus to take place in most female children. Although the mass vaccination program should not be terminated until results are known, laws requiring rubella vaccination should be suspended until such procedures are shown to be in the best interests of the entire community. (No refs.) - *S. Glinsky.*

Wistar Institute
Philadelphia, Pennsylvania

2640 **ABRAMSON, JOSEPH H.; SACKS, THEODORE C.; FLUG, DOROTHY; ELISHKOVSKY, RUTH; & COHEN, RIVKA.** Bacteriuria and hemoglobin levels in pregnancy. *Journal of the Amer-*

ican Medical Association, 215(10): 1631-1637, 1971.

Among women attending 3 antenatal clinics in Jerusalem, Israel, an association was observed between significant bacteriuria and low hemoglobin values. The Ss were 652 pregnant women upon whom urine tests were performed in early and late pregnancy. Significant bacteriuria was found in 13 women, probable significant bacteriuria in 18, and possible significant bacteriuria in 20. Those with definite or probable bacteriuria also tended to have lower hemoglobin values than nonbacteriuric women. This association did not seem to be caused solely by effects connected with age, parity, education, social class, region of birth, date of immigration, or diet. The findings suggest the value of screening programs for bacteriuria in antenatal clinics and of urine cultures in women with anemia of pregnancy. (15 refs.) - *S. Glinsky.*

Hebrew University-Hadassah Medical School
Jerusalem, Israel

2641 **HERRMANN, KENNETH L.; WENDE, REUBEN L.; & WITTE, JOHN J.** Rubella immunization with HPV-77 DE_5 vaccine during infancy. *American Journal of Diseases of Children,* 121(6): 474-476, 1971.

In addition to passively acquired maternal antibody other factors strongly influenced the immunologic response of 137 infants between 6 and 12 months of age to live rubella vaccine. Testing of serial blood specimens taken from 10 infants from birth to 6 months of age resulted in the definition of the half-life of rubella H1 antibody as 30 days. Data confirmed that rubella antibody is rarely detectable after 6 months of age. No rubella H1 antibody was detectable in any study participant at the time of vaccine inoculation. Yet only 69% of the infants under 10 months of age seroconverted after rubella vaccine inoculation, while over 90% of those 10 months of age and older responded positively. There was no correlation between failure to seroconvert and levels of passively acquired rubella antibody at birth. These results suggest that other age related factors strongly influence the infant immunologic responses to live rubella vaccine. (5 refs.) - *S. Glinsky.*

Health Services and Mental Health Administration
Atlanta, Georgia 30333

2642 **SIEGEL, MORRIS; FUERST, HAROLD T.; & GUINEE, VINCENT F.** Rubella epidemicity and embryopathy. *American Journal of Diseases of Children,* 121(6): 469-473, 1971.

In a long-term prospective study of maternal rubella the incidence of major malformations was higher in the extensive outbreak of 1964 than in the period 1952 to 1963, when the disease was less widespread. The Ss were 381 cases of maternal rubella who were observed to age 5 of the offspring. Malformations occurred in 54.2% of the 1964 cases as compared with 30.6% for the prior period in cases of rubella acquired during the first trimester. The difference was mainly due to a significantly higher incidence of congenital cataracts, deafness, and cardiac abnormalities in 1964 than in the previous 7 years. The findings may be attributed either to the increased teratogenicity of the 1964 rubella virus or to greater accuracy in clinical diagnosis of maternal rubella during that period. An explanation based on a biological change in the virus is consistent with the almost pandemic occurrence of rubella in 1964. (13 refs.) - *S. Glinsky.*

State University of New York
College of Medicine
New York, New York

2643 **McELRATH, THOMAS G.; & ASCARI, WILLIAM Q.** Prevention of Rh immunization after transfusion with Rh-positive blood. *American Journal of Obstetrics and Gynecology,* 110(3): 309-310, 1971.

Rh-immune globulin was efficacious in preventing Rh immunization after transfusion of Rh-positive blood into an Rh-negative woman. A nulliparous Rh-negative woman was transfused with 500 ml of Rh-positive blood after acute blood loss associated with an incomplete abortion. Within 72 hours after transfusion 20 vials of Rh-immune globulin were given to her in divided doses. This resulted in the development of a passive anti-Rh titer of 1:16 and the disappearance of the transfused Rh-positive cells from her blood over a 12 day period. The patient experienced no adverse reaction, and an 8 month follow-up indicated no evidence of active Rh immunization. The ultimate test will be an uncomplicated subsequent pregnancy. (2 refs.) - *S. Glinsky.*

Hunterdon Medical Center
Flemington, New Jersey

2644 OH, WILLIAM; ARBIT, JACK; BLONSKY, E. RICHARD; & CASSELL, SYLVIA. Neurologic and psychometric follow-up study of Rh-erythroblastotic infants requiring intrauterine blood transfusion. *American Journal of Obstetrics and Gynecology*, 110(3): 330-335, 1971.

Neurologic and psychometric follow-up study of Rh-erythroblastotic infants who received intrauterine blood transfusions suggests a normal developmental performance at a later age. The Ss were 10 infants surviving from severe Rh erythroblastosis requiring intrauterine blood transfusion. Controls were 10 normal infants matched for birth weight, gestational age, sex, race, home environment conditions, and age at the time of examination. Neurologic, psychometric, and EEG findings indicated no differences between the Rh survivors and controls. (15 refs.) - *S. Glinsky.*

Michael Reese Hospital and Medical Center
Chicago, Illinois

2645 CLAYTON, EVERETT M., JR.; WALLER, DAVID H.; & FOSTER, ELINOR B. Antepartum and postpartum use of passive Rh antibodies. *American Journal of Obstetrics and Gynecology*, 107(5): 704-707, 1970.

In a small number of patients postpartum intravenous administration of Rh hyperimmune human plasma is effective in preventing Rh sensitization. High-titer human plasma was prepared from 2 donors and intravenously given postpartum to 145 patients. Rh sensitization occurred in all cases, and neither the 2 donors nor patients showed any signs of hepatitis. An effective amount of antibody given to protect against Rh sensitization was safe and harmless to the unborn fetus. There is no value in the administration of passive Rh antibodies for the suppression of the secondary response in patients who are already sensitized. (5 refs.) - *S. Glinsky.*

Saint Thomas Hospital
Nashville, Tennessee

MEDICAL ASPECTS — Etiologic Groupings

Trauma or physical agents

2646 The schoolchild with a squint. *Lancet*, 1(7713): 1342, 1971. (Editorial)

An investigation of the significance of squinting at age 7 years in a survey grouping of over 10,000 English children who have been medically followed since birth reveals an apparent association between this condition and other signs of cerebral dysfunction. Of the 3.5% who exhibited a squint, 1.6% were found to be SMR or to have cerebral palsy, as compared to 0.3% of controls. An additional 1.2% were found to be EMR, as compared to 0.4% of controls. When this cohort was excluded from a further study to determine the effects of squinting on educational achievement, evidence supported a relationship between this condition and poor reading performance, even in the absence of other signs of mild cerebral dysfunction, such as clumsiness, hyperactivity, and general maladjustment. Clearly the presence of squinting in a child should alert the physician or teacher to the possible existence of other disabilities, particularly if birthweight was low. (3 refs.) - *N. Mize.*

2647 TEC, LEON. Amphetamines in hyperkinetic children. *Journal of the American Medical Association*, 216(11): 1864, 1971. (Letter)

To date, there is no firm evidence to suggest that the administering of amphetamines or methylphenidate to hyperkinetic children conditions them to rely on drugs generally to alleviate unpleasant situations. Most recent experience would tend to attribute a biological rather than an environmental etiology to the development of the hyperkinetic syndrome, suggesting that the oft-noted signs of parental mismanagement could be the negative result of parental frustration at trying to raise a hyperactive child, and not the cause of this condition. (No refs.) - *N. Mize.*

No address

2648 LEVY, HAROLD B. Amphetamines in hyperkinetic children. *Journal of the*

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American Medical Association, 216(11): 1864-1865, 1971. (Letter)

The suggestion that administration of medication to hyperkinetic children suffering from minimal brain dysfunction may predispose to later drug addiction is unfounded. Instead it is more commonly the case that as such children grow older they show resentment toward their earlier dependence on medication and tend to avoid drugs of all types. Neither is it true that minimal brain dysfunction is linked significantly to lack of parental authority and love; instead, the most current research links this condition with faulty norepinephrine metabolism. Since the administering of methylphenidate, especially, has been shown to improve markedly the behavior and scholastic performance of the hyperkinetic child, its continued judicious use may well be a most important contribution to preventive psychiatry. (9 refs.) - *N. Mize.*

No address

2649 **BELMONT, IRA; & HANDLER, ALAN.** Delayed information processing and judgment of temporal order following cerebral damage. *Journal of Nervous and Mental Disease*, 152(5): 353-361, 1971.

Comparison of 21 hospitalized left hemiplegic patients and 21 non-neurologically impaired Ss matched for age, sex, education, socioeconomic background, and length of hospital residence tends to support the view that damage to the central nervous system leads to a retardation in speed of information processing. All Ss were required to judge the temporal order of 2 successively presented auditory stimuli when first 1 ear and then the other was stimulated. Findings strengthened the inertia hypothesis, with hemiplegic patients tending to report that the ear related to the affected hemisphere was stimulated last, independently of the real order of stimulation, while normal Ss responded accurately. Further studies on other brain damaged populations, particularly those with left cerebral damage, are needed to lend additional support to these findings. (13 refs.) - *N. Mize.*

Albert Einstein College of Medicine
New York, New York 10461

2650 **SUNDELL, HAKAN; GARROTT, JOHN; BLANKENSHIP, WILLARD J.; SHEPARD, FRANK M.; & *STAHLMAN, MILDRED T.** Studies on infants with type II respiratory distress syndrome. *Journal of Pediatrics*, 78(5): 754-764, 1971.

Studies of a group of infants with clinical, biochemical, and physiologic similarities have indicated that their disorder, type II respiratory distress syndrome, has initial clinical symptoms similar to but distinguishable from clinical hyaline membrane disease (CHMD). Of a patient population of 36 newborn infants with respiratory distress (29 male and 7 female) 29 were available for follow-up evaluation at periods ranging from one to 7 years, with a mean age at last testing of 3.2 years. Initial and early characteristics and symptoms include prematurity, a frequent history of maternal sedation or a cesarean delivery, mild depression, and respiratory symptoms (grunting, chest-wall retraction, tachypnea, flaring of the alae nasal). The infants were amenable to hyperoxygenation and had early evidence of mildly to moderately severe metabolic and respiratory acidosis. Typical radiographic findings differed strikingly from those of CHMD, with the diffuse reticulogranular pattern characteristic of CHMD never seen in these Ss. Follow-up evaluations showed that prognosis for this type of respiratory distress is good. Symptoms of the respiratory distress subside in 3-7 days after birth; pulmonary symptoms and radiologic abnormalities do not persist; and normal motor and mental development ensue. (11 refs.) - *M. S. Fish.*

*Vanderbilt University School of Medicine
Nashville, Tennessee 37203

2651 **WARREN, RICHARD J.; KARDUCK, WILLIAM A.; BUSSARATID, SAMPORN; STEWART, MARK A.; & SLY, WILLIAM S.** The hyperactive child syndrome. *Archives of General Psychiatry*, 24(2): 161-162, 1971.

Chromosome analysis was done on 96 Ss (82 boys and 14 girls) to determine whether chromosome anomalies had a part in the origin of the hyperactive child syndrome. Two cytological procedures were used: sex chromatin survey and a complete karyotypic analysis of 20 boys and 3

girls. No evidence for an abnormal number of X chromosomes was found in any of the patients, nor did the karyotype analysis reveal any abnormality of chromosome number or morphology. It was concluded that chromosome abnormality is not a major cause of the hyperactive child syndrome. (15 refs.) - *T. C. Turner.*

Mailman Center for Child Development
Miami, Florida 33152

2652 **REYNOLDS, E. O. R.** Effect of alterations in mechanical ventilator settings on pulmonary gas exchange in hyaline membrane disease. *Archives of Disease in Childhood*, 46(246): 152-159, 1971.

Six infants undergoing mechanical ventilation on a Bennet ventilator because of hyaline membrane disease were exposed to alterations in peak airway pressure, respiratory frequency, and inspiration:expiration ratio in an attempt to affect arterial blood gas tensions, blood pressure, and right-to-left shunt. Slow respiratory frequency (30 breaths per minute) and high peak airway pressure (25 cm H_{20}) produced optimal arterial oxygen tension. This could be improved by increasing the peak airway pressure or increasing the inspiration:expiration ratio; however, long term effects of these procedures have not been determined. Increasing the inspiration:expiration ratio did decrease the right-to-left shunt without altering arterial blood pressure. (33 refs.) - *E. L. Rowan.*

University College Hospital
London W.C. 1, England

2653 **WEISS, GABRIELLE; MINDE, KLAUS; *WERRY, JOHN S.; DOUGLAS, VIRGINIA; & NEMETH, ELIZABETH.** Studies on the hyperactive child. VIII. Five-year follow-up. *Archives of General Psychiatry*, 24(5): 409-414, 1971.

Five year follow-up data on hyperactive children indicated that although hyperactivity appeared to decrease with age, other major handicaps persisted. Selective criteria for inclusion in the original study for the 64 Ss were: CA between 6 and 13 years; Full Scale WISC IQ greater than 84; absence of major brain damage or dysfunction; non-psychotic; and residing at home with at least 1 parent. 24 Ss were initially diagnosed as hyperactive and 40 were described as restless. Areas evaluated at follow-up included behavior, social adaptation, academic function, cognitive and motor function, neurophysiological status, and family status. The children did not appear to outgrow restless behavior entirely but seemed to express it in ways that were less gross and disturbing. They exhibited more restless behavior, were more aggressive, and performed more antisocial acts than a control group of normal peers. As a group they were emotionally immature, found it difficult to maintain goals, had a poor self image, and expressed feelings of hopelessness. 80% had poor academic records, 70% had repeated at least 1 grade, and 10% had been placed in EMR classes for varying lengths of time. Children with initially high ratings on aggression had a higher incidence of later antisocial behavior. (14 refs.) - *J. K. Wyatt.*

*University of Auckland
School of Medicine
Auckland, New Zealand

2654 **BAX, M.** The larger half. *Developmental Medicine and Child Neurology*, 13(2): 135-136, 1971.

Learning and behavior problems of the school-age child may occupy a greater portion of contemporary pediatric practice than is generally assumed. The casual use of drug treatment for such overactivity is to be abhorred. The pediatrician must approach the difficulty in terms of interaction of child, parents, peers, and teachers and reserve drugs for the few true cases of organic hyperactivity. The management of overactivity calls upon more resources than the physician is accustomed to providing. (2 refs.) - *E. L. Rowan.*

No address

2655 **PELC, ROBERT E.** Advancement along a complexity gradient in perceptual level and brain damage. *Perceptual and Motor Skills*, 32(1): 251-254, 1971.

A comparison of brain-damaged subjects (N=40), psychiatric patients (N=40), and normal controls

(N=40) on performance on the Goodenough Draw-A-Person Test and the Bender-Gestalt revealed less complexity in the work of brain-damaged subjects. They included fewer details on the Goodenough and made greater deviations on the Bender. The low level of complexity reflected impaired perceptual ability in the brain-damaged rather than cognition, as their intellectual functioning was not different from that of controls. (9 refs.) - *E. L. Rowan.*

Fitzsimmons General Hospital
Denver, Colorado

2656 MILLER, JAMES A. New approaches to preventing brain damage during asphyxia. *American Journal of Obstetrics and Gynecology,* 110(8): 1125-1133, 1971.

Several methods of combating asphyxia in neonates have been proven effective under carefully controlled conditions. Survival times for neonates during asphyxia have been found to be longest not at 37°C but at 15°C body temperature in all but 2 species tested (guinea pig and pig). For both kittens and puppies, cooling initiated after the onset of asphyxia enables neonates to recover spontaneously from exposures which are lethal for warm littermates. Hypothermia has also been shown to protect the asphyxiated brain of newborn rhesus monkeys from damage. Hypothermia reduces biochemical decay during asphyxia as measured by 0_2 uptake, serum K^+, glucose, lactate, pyruvate, pH, and P_{CO_2}. The most striking effects of hypothermia during asphyxia are on cardiovascular function, with improvements noted in the electrocardiogram, blood pressure, and pulse pressure. In combination with tris hydroxymethyl aminomethane (THAM) and glucose, hypothermia prolonged the time of last gasp in puppies by 69%. On the basis of early human trials, it has been recommended that human neonates with Apgar scores of 3 or less be cooled immediately after the airways have been cleared and the lungs have been expanded. The use of intra-arterial transfusion for resuscitation of the asphyxiated newborn infant has been largely limited. Intravenous transfusion is ineffective in deep human asphyxia, when cardiac efficiency is impaired. (27 refs.) - *B. J. Grylack.*

Tulane University
New Orleans, Louisiana

2657 WESTIN, BJORN. Infant resuscitation and prevention of mental retardation. *American Journal of Obstetrics and Gynecology,* 110(8): 1134-1138, 1971.

An active approach to the patient in labor has resulted in improvement in perinatal statistics during the last decade in Sweden. In infants born with asphyxia, aeration of the lungs appears to be needed to decrease pulmonary vascular resistance. Since it is only between inflations that pulmonary blood flow is increased during resuscitation, care must be taken to perform inflations at a slow rate and with short duration, under close observation of the heart rate. In the choice between infusion with base and transfusion with oxygenation, the latter is preferred because it offers both buffer and oxygen in addition to plasma expander. Contrary to practice in many centers, exposure of the deeply asphyxiated infant to cold regularly results in an immediate increase in heart rate and a rapid increase in Apgar scores. There has been a report of a series in which 20 of 22 infants with a mean Apgar score of 2 before cooling survived. Another series of 60 infants was treated with cooling, and the 9 children examined again at age 10 showed normal development. Cooling of asphyxic neonates may serve to prevent loss of mental function. (32 refs.) - *B. J. Grylack.*

Danderyds Hospital
Danderyds, Sweden

2658 BIERBAUER, ELAINE. The educationally handicapped child. *Journal of the American Medical Association,* 217(4): 482, 1971. (Letter)

The physician's difficulty in counseling parents in the home management and education of minimally brain-damaged children is presented. Physicians must know what public and private resources are available in the community to aid the children, and must be able to inform the parents about special education programs, speech and hearing clinics, remedial reading clinics and so forth. Only by providing parents with an understanding of the disorder and information about community resources can the physician claim to be giving total care to the minimally brain-damaged child. (No refs.) - *T. C. Turner.*

No address

2659 **DOBIE, ROBERT A.; & SIMMONS, F. BLAIR.** A dichotic threshold test: Normal and brain-damaged subjects. *Journal of Speech and Hearing Research*, 14(1): 71-81, 1971.

Testing was administered to 33 normal right-handed and left-handed individuals and to a group of 10 patients with unilateral cerebral lesions in order to develop and assess a single-report paradigm, according to which Ss attend to and report the items presented to 1 ear and ignore the items presented to the other ear. Nonsense syllables served as test items. Ten tokens of each were recorded monaurally; oscilloscope records of these were photographed, and 1 token of each class was selected on the basis of burst duration. The average dichotic threshold difference for right-handed Ss was shown to differ significantly, and in the expected direction, from the same score obtained for left-handed Ss. Ss in the right-handed group performed better with their right ears in this task. Left-handed Ss performed slightly better with their left ears, but this comparison was not at all statistically significant. Six of the brain-damaged Ss had nondominant cerebral lesions. The finding that 4 of them showed markedly superior performance by the ear opposite the intact dominant hemisphere suggests that there is a center in the nondominant hemisphere which is important as a relay for speech information from the contralateral ear. (8 refs.) - *B. J. Grylack.*

Stanford University
Stanford, California

2660 **KROP, HARRY.** Modification of hyperactive behavior of a brain-damaged, emotionally disturbed child. *American Institute for Mental Studies Training School Bulletin*, 68(1): 49-54, 1971.

The effectiveness of applying operant techniques to shape the attending behavior of a MR brain-injured, emotionally disturbed boy (CA 8 years) is demonstrated. After baseline observations were made to record when hyperactive behavior was or was not occurring, the occupational therapist gave primary and secondary reinforcements to the S when he was showing nonhyperactive behavior. The S adjusted within a week to the new program and his behavior improved markedly. A follow-up study indicated that this behavior was maintained. (6 refs.) - *T. C. Turner.*

Gainesville Veterans Administration Hospital
Gainesville, Florida

2661 **LITTLETON, ARTHUR C.; & DAVIS, FRANCES.** The mystique of brain injury. *American Institute for Mental Studies Training School Bulletin*, 68(1): 15-22, 1971.

A brief review is made of some of the central issues requisite to an understanding of the concept of brain injury. Covered are: pathological as opposed to behavioral injury; brain injury and learning disabilities; the minimally brain-injured person as distinguished from the MR; the high incidence of prenatal, paranatal, and postnatal brain damage; the perceptual problems of the brain-injured child; the value of certain drugs as a treatment approach; the education of the brain-injured. Some of the complex issues regarding terminology and several behavioral implications are also discussed. (15 refs.) - *T. C. Turner.*

University of Missouri
St. Louis, Missouri

2662 Checklist for the child with brain dysfunction. *Expectations*, 1(2): 3, 1971.

Parents and teachers can identify a child with a perceptual handicap or brain dysfunction by looking for these characteristics: 1) Low achievement in some areas versus high in others; 2) Poor eye-hand coordination and distortion in drawing of simple shapes; 3) Able to follow oral instructions much better than written; 4) slow reading ability, mirror writing, reversal of letters, words, numbers; 5) Difficulty in comprehending meaning of numbers and in telling time; 6) Poor phonetic discrimination, confusion of letters or words; 7) Short attention span, restless and impulsive in class, hyperactive; 8) Asks same questions many times or repeats words; 9) Low frustration level and performance in peer relationships and sports; 10) Fluctuating performance levels; is slow at or cannot finish a task; 11) Hyperactive; 12) Has great difficulty in choosing, when given several choices. Once these children

are identified as having learning disabilities (not retarded, lazy, or willfully disobedient) they can be placed in programs which will help them improve. (No refs.) - *T. C. Turner.*

2663 **PETERS, ANN.** "absurd to deny..." *Expectations*, 1(2): 4, 1971.

An argument is made for the necessity of allowing treatments without a thoroughly understood conceptual basis to be used for disabilities of brain dysfunction even though they are not effective on all patients. If the treatment proves successful in some cases, then it should be utilized, perhaps with improvements and refinements, even though it is not supported with a scientific explanation. Since parents are willing to accept new treatments, the professionals in the field should also be willing to try something having a chance of success. (No refs.) - *T. C. Turner.*

2664 **KASPAR, JOSEPH C.; MILLICHAP, J. GORDON; BACKUS, RENO; CHILD, DAVID; & SCHULMAN, JEROME L.** A study of the relationship between neurological evidence of brain damage in children and activity and distractibility. *Journal of Consulting and Clinical Psychology*, 36(3): 329-337, 1971.

The possibility of a relationship between brain injury and behavioral measures of activity and distractibility was tested in 24 males and 12 females between the ages of 5 and 8 who were of normal intelligence but who were diagnosed as damaged on the basis of neurological examination and in an equal number of control Ss matched for sex. Control Ss were found to be significantly more intelligent than experimental Ss, but the average IQs of both groups were within the normal range. In a free situation, the experimental group was not significantly more active than the control group, but in a structured task the damaged Ss produced approximately twice as much activity as did the control Ss. These results support the hypothesis that the effect of brain damage is not to increase the amount of activity a child generates in a free situation but to make it more difficult for the child to reduce his activity level in situations in which such a reduction is required or anticipated. Three of 4 distractibility tests differentiated the 2 groups beyond the .001 level, with more brain-injured than control Ss shown to be distractible. These data substantiate the premise that activity levels and distractibility are increased in Ss with clinical evidence of brain dysfunction. (16 refs.) - *B. J. Grylack.*

Children's Memorial Hospital
Chicago, Illinois 60614

2665 **DeVIVO, DARRYL C.; & *DODGE, PHILIP R.** The critically ill child: Diagnosis and management of head injury. *Pediatrics*, 48(1): 129-138, 1971.

Head injury ranks high among the causes of death and disabilities in childhood. Even though the diagnosis of head injury seems obvious, a detailed present and past history are essential to reveal such conditions as drug allergies, hemophilia, diabetes mellitus, and epilepsy, which may bear significantly on the course of the child. After a thorough clinical evaluation, skull and other roentgenograms are usually indicated, especially if the patient lost consciousness following the injury. The majority of infants and children who have not lost consciousness following head injury can be cared for by their parents. Those patients who have focal or diffuse neurological disturbances and all patients who have been rendered unconscious for a period of time with or without an associated skull fracture should be hospitalized until the condition is stable and the neurological signs abate. Much of the management of the patient with acute head injury has been arrived at empirically, and many of the suggested clinical regimens, including the choice and dose of various therapeutic agents, are arbitrary. Clinical and basic research are needed in regard to the common problems associated with acute head injury. (19 refs.) - *B. J. Grylack.*

*500 S. Kingshighway
St. Louis, Missouri 63110

2666 **KLEIN, MICHAEL; & STERN, LEO.** Low birth weight and the battered child syndrome. *American Journal of Diseases of Children*, 122(1): 15-18, 1971.

A contributing factor to battered child syndrome (BCS) may be low birth weight associated with

premature birth, resulting in a separation between mother and infant during a neonatal period of intensive care. The records of all BCS patients (51 Ss) from 1960 to 1969 at Montreal Children's Hospital were reviewed and categorized as to age at diagnosis, sex, birth weight, gestation period, maternal age, type of battering, newborn problems, evidence of maternal-child separation in neonatal period, MR, deprivation in the maternal background, and outcome. Twelve of the Ss were underweight at birth (less than 2,500 gm), representing 23.5% of the BCSs studied. The expected proportion of low-weight births in the population, based on the Quebec perinatal figures, is 7-8%. Nine of the 12 low-birth-weight infants had major neonatal problems, and the hospital stay ranged up to 80 days (mean, 41.4 days). Six had psychomotor retardation prior to battering; 10 came from deprived home environments in which the mother had usually suffered maternal and environmental deprivation as a child. There was a high incidence of parental alcoholism, unemployment, extreme poverty, and social disorganization. The incidence of low-weight births among the indigent is somewhat higher (1-2%) than the average, but not sufficiently higher to provide a social explanation for the high proportion of low-birth-weight infants in the BCS sample. As a possible remedy, it is suggested that mothers be brought into the hospital nurseries during the neonatal intensive care period to handle their infants. This also would give the staff opportunity to observe the mother with her child and provide clues to the mother's ability to care for and cope with what may be a difficult child. (11 refs.) - *S. M. Rucker.*

Strong Memorial Hospital
Rochester, New York 14620

2667 VEGA, ARTHUR; & PARSONS, OSCAR A. Lateralized brain damage and differential psychological effects: Reply to Dr. Woo-Sam. *Perceptual and Motor Skills,* 33(1): 269-270, 1971.

A critique which questions the finding of differential effects of left-hemisphere as compared to right-hemisphere brain damage on the Wechsler Vocabulary and Block Design subtests is largely unjustified. The critic overlooks the weight of evidence favoring the differential effects of left- and right-hemisphere lesions and misinterprets a study which emphasizes that differential performance on the Wechsler scale is to be expected. In addition, his criticism of the use of age-corrected scores contradicts the recommendation made by Wechsler himself that this technique be employed, generally, in any pattern analysis which utilizes deviation scores. Moreover, the critic's own statistical treatment of the data is inappropriate, and it is difficult to determine exactly what position his negative results support, since they do not even illustrate any deterioration of the Block Design subtest of the Performance scale as a function of either right- or left-hemisphere pathology. (3 refs.) - *B. J. Grylack.*

University of Oklahoma Medical Center
Norman, Oklahoma

2668 WOO-SAM, JAMES. Lateralized brain damage and differential psychological effects: Parsons, et al., re-examined. *Perceptual and Motor Skills,* 33(1): 259-262, 1971.

A study by Parsons and colleagues of the differential effects of lateralized brain damage on the Wechsler scales was reanalyzed in order to review their conclusion that left-hemisphere damage was associated with a Block Design score significantly higher than the Vocabulary score, while the converse was held to be true when the right hemisphere was damaged. Twenty-one right-hemisphere-damaged and 15 left-hemisphere-damaged head-injured veterans were used in a further analysis to test those reported differences. The use of age-corrected scale scores by Parsons tended to exaggerate or attenuate inappropriately the differences in laterality and, therefore, to weaken the concluded relationship between psychological effects and laterality. Age-compensated scores for every age group would allow for comparisons of the direction of differences across the various age ranges. The new analysis, which was based upon Ss with equivalent CAs, showed no differential resulting psychological effects attributable to the location of injury. (8 refs.) - *B. J. Grylack.*

Rancho Los Amigos Hospital
Downey, California

2669 SALMON, JAMES H.; & TIMPERMAN, ALBERT L. Cerebral blood flow in

posttraumatic encephalopathy: The effect of ventriculoatrial shunt. *Neurology*, 21(1): 33-42, 1971.

Cerebral blood flow was determined on 21 occasions in 12 patients with chronic posttraumatic encephalopathy by use of the intracarotid xenon 133 technique. The cerebral blood flow of the gray matter (CBF_G) was decreased in all patients, and little correlation was found between the CBF_G and the severity of clinical disability. The cerebral blood flow of white matter (CBF_W) varied from 45 to 130% of normal. CBF_G is ordinarily about 4 times greater than CBF_W. In the chronically demented patients, CBF_G was only twice as great as CBF_W, a finding which may indicate that blood was being diverted from the gray matter vessels through the white matter vessels. The parameter that correlated best with the duration and severity of injury was the relative weight of gray matter (W_g). The cerebrovascular resistance of the gray matter (CVR_G) varied inversely with CBF. The frontal cerebral mantle was measured by air encephalography and was found to vary with W_G. A ventriculoatrial shunt was inserted in 6 patients. Cortical CBF did improve with intracranial hypotension, and this increase in CBF_G was associated with clinical improvement. Since the lowering of intracranial pressure led to increased cerebral blood flow, further research will prove valuable in that area. (17 refs.) - *B. J. Grylack.*

Veterans Administration Hospital
Cincinnati, Ohio 45220

2670 **TOWBIN, ABRAHAM.** Organic causes of minimal brain dysfunction. Perinatal origin of minimal cerebral lesions. *Journal of the American Medical Association*, 217(9): 1207-1214, 1971.

The human fetus and newborn frequently suffer irreversible hypoxic injuries which lead to subsequent impairment of neurologic function. Clinically, in addition to the 3 commonly recognized sequels of neonatal brain damage--MR, cerebral palsy, and epilepsy—attention has been focused on behavioral disorders. Data collected on 600 neonatal cases have indicated that the 4 main forms of central nervous system damage are subdural hemorrhage, due to traumatic dural-venous tears; spinal cord and brain stem damage due to mechanical injury; hypoxic damage to the deep cerebral structures, occurring predominantly in the premature, and hypoxic damage to the cerebral cortex, occurring mainly in the mature fetus and newborn. Evidence of minimal brain dysfunction resulting from neonatal damage is, of course, more subtle than gross forms of dysfunction and may not be recognized until late childhood or early adulthood. (17 refs.) - *B. J. Grylack.*

Danvers State Hospital
P. O. Hathorne, Massachusetts 01937

2671 **SCHULTE, F. J.** Current concepts in minimal brain dysfunction. *Journal of the American Medical Association*, 217(9): 1237-1238, 1971.

Current thinking in the area of minimal brain dysfunction tends to place less emphasis on the classical etiologies of birth trauma and hypoxia as factors to be considered in perinatal minimal brain damage. More attention is being focused on inherited metabolic peculiarities, intrauterine and extrauterine malnutrition, and aspects of the early postnatal environment such as light, temperature, and handling. Some early evidence of minimal dysfunction may be found through the sophisticated evaluation of the motor behavior of the infant and the application of statistical analysis, quantitative analysis of the neonatal EEG with pattern recognition and automatic spectrum analysis, and observation of the rhythmic behavior of the sleep/wakefulness cycle. Insidious consequences of minimal brain damage include school problems, mild developmental retardation, minor motor incapabilities, and behavioral anomalies. (12 refs.) - *B. J. Grylack.*

University of Gottingen
Gottingen, Germany

2672 **HARRIS, T. R.** Respiration monitor for infants with tracheostomy. *Lancet*, 2(7724): 583-584, 1971.

A newly developed dependable and convenient monitoring system for infants with chronic tracheostomies (complications, such as mucus obstruction of the inner cannula, require continual supervision) gives advance warning of occlusion. Sensor and alarm portions of the system are

from the Lewin apnea-alarm mattress. On inspiration or expiration, air rushing through the tracheostomy opening sets up a Venturi effect in connector tubes, drawing clean, dry, cooling air into an open manifold. Experience in using the device on 2 Ss is reported. (4 refs.) - *B. Berman.*

University of Arizona College of Medicine
Tucson, Arizona 85721

2673 TURNER, E. J.; & ARNOLD, L. EUGENE. Mood-altering drugs (continued). *Pediatrics,* 48(3): 496-497, 1971. (Letter)

Various opinions have been recorded in response to an article concerning the use of mood-altering drugs in children and the problems surrounding this practice. It has been suggested that some physical punishment administered by the teacher to the hyperactive child might be better than continual administration of antihyperactive drugs. In contrast, it has been emphasized that the long-range effects of leaving a hyperkinetic syndrome untreated should be of greater concern than the long-range effects of stimulant medication. (4 refs.) - *B. J. Grylack.*

10830 Jasper Avenue
Edmonton, Alberta, Canada

2674 FISH, BARBARA. The "one child, one drug" myth of stimulants in hyperkinesis. *Archives of General Psychiatry,* 25(3): 193-203, 1971.

There are many types of hyperactive children, and it is not true that stimulants are good for all hyperactive children or that there is "one drug for one child." Treatment of 500-600 children with dextroamphetamine was ineffective not only with the overtly schizophrenic but also with those who resembled psychotics in their excitement, irritability, and impulsive hyperactivity. Most of the so-called contradictions in studies on stimulants in children are artifacts caused by applying different terms to the same children. Confusion is compounded when "hyperactive reaction" is used interchangeably with "minimal brain dysfunction." Many nonhyperactive children respond to stimulants, as do some with minimal brain dysfunction. We must distinguish between diagnoses defining the type of total personality disorder and the terms relating to major developmental symptoms—both groups are vital in diagnosing children's disorders. (27 refs.) - *B. Berman.*

New York University School of Medicine
New York, New York

MEDICAL ASPECTS — Etiologic Groupings

Disease or disorders of metabolism, growth, or nutrition

2675 Diagnosis before birth. *Medical World News,* 12(19): 64F, 1971.

For women at risk of Niemann-Pick disease, transabdominal amniocentesis is now being used to identify *in utero* the fetus affected with this inherited enzymatic disorder. If cultured fetal cells show the greatly reduced sphingomyelinase activity characteristic of this condition, therapeutic abortion may be offered. (No refs.) - *N. Mize.*

2676 Hurler's syndrome "factor" found in human serum. *Journal of the American Medical Association,* 215(5): 716, 1971.

New possibilities in the diagnosis and treatment of Hurler's syndrome have been opened up with the recent discovery of a "corrective factor" for this disease in normal human serum. Groups of $S^{35}O_4$ - labeled Hurler cells from an afflicted 6-year-old boy were treated with media exposed to either normal human cells or other Hurler cells. On analysis, the amount of labeled mucopolysaccharides remaining in the group treated with normal cell media was less than 1/2 that in the solution treated with media exposed to other Hurler cells. Serum from pregnant women is particularly effective in promoting this mucopolysaccharide degradation, though that from non-pregnant women also contains the corrective factor, but in lesser amounts. Further research in this area will greatly aid early diagnosis and therapy in cases of suspected metabolic defects of the Hurler variety. (No refs.) - *N. Mize.*

2677 **HSIA, DAVID YI-YUNG.** A critical evaluation of PKU screening. *Hospital Practice*, 6(4): 101-104, 109-112, 1971.

Recent experience with large scale screening for phenylketonuria (PKU), a program which first dates from the mid-1950's in England, has pointed up the need for a critical evaluation, on a scientific and socioeconomic basis, of the value and limits of routine newborn screening for inborn errors of metabolism. Research stimulated by refinements in testing procedures, from the FeCl diaper technique to the Guthrie test, has revealed a wide range of variant forms of hyperphenylalanemia, with a continuous distribution of PKU values ranging from the 6 mg% defined by the Guthrie test as a presumptive positive to 80 mg% and more. The variant and milder forms, constituting an estimated 1/3 to 1/2 of all positive test results, exhibit none of the clinical effects of classic PKU, and affected children appear to be of normal intelligence. Many of these, however, have been unnecessarily, and perhaps dangerously, treated with special diets. The best compromise between the overall costs of the mass screening and treatment program, and the number of positive MR-associated PKU cases treatable by early diet modifications who are so identified, lies in redoubling research efforts to enable simultaneous screening of several genetic diseases in the neonate population. Priority should also be given to screening those especially susceptible population groups. Already some states which had earlier enacted compulsory PKU screening laws are eliminating the program, because of expense, in low frequency population areas. The epoch of mass screening for hereditary metabolic defects in the newborn has only just begun, and the experience with PKU programs must be carefully evaluated in public forums to minimize future problems which might accompany program expansion. (No refs.) - *N. Mize.*

Loyola University
Stritch School of Medicine
Maywood, Illinois

2678 Mysterious plasma 'factors' soften 'irreversible' gargoylism. *Medical World News*, 12(15): 28, 1971.

Marked improvement in the clinical state of young Hurler and Hunter syndrome victims has followed the experimental infusion of large quantities of normal human plasma. Normal plasma apparently contains "corrective factors" for mucopolysaccharidosis, though the specific enzyme or enzyme component involved has yet to be isolated biochemically. This discovery is the first evidence that such previously uncorrectable inborn errors of metabolism may yield to enzyme replacement, previous attempts to modify the disease course of gargoylism with vitamins, hyperbaric oxygen, and longterm corticosteroid therapy, have met with little success. (No refs.) - *N. Mize.*

2679 **SOLCHER, H.; & WOLFF, D.** Spatinfantile amaurotisch Idiotie mit ungewohnlich langem Verlauf (Late infantile amaurotic idiocy of unusually protracted course). *Acta Neuropathologica*, 13(4): 338-349, 1969.

A 25-year-old female patient exhibited characteristics of late infantile amaurotic idiocy of the ganglioside storage type. After a normal birth and early childhood, cerebral convulsions without fever began at 3 years of age. Subsequently she became incapable of speech, hearing, and intellectual performance, and her personality deteriorated. In her seventh year the lower extremities became progressively ataxic, and by 17 years of age the patient had increased ataxia of the upper extremities and the trunk, which caused her to become bedridden. Marked impairment of vision, preterminal disorders of swallowing and breathing, and myoclonic convulsions of the right extremities occurred. Neuropathological investigation of the brain showed cellular storage of lipid material and demyelination accentuated in the cerebellum, pons, olivar, and optical system. A clinical history of 23 years is uncommon for the late infantile amaurotic idiocy which the findings revealed. Due to the acute onset of the disease and the morphological findings this case cannot be classified as an aged late case as described by Hallervoden. (16 refs.) - *S. Glinsky.*

Universitats-Nervenklinik
355 Marburg a.d. Lahn, Germany

2680 **HOWELL, R. RODNEY; KABACK, MICHAEL M., & BROWN, BARBARA I.** Type IV glycogen storage disease: Branching enzyme deficiency in skin

fibroblasts and possible heterozygote detection. *Journal of Pediatrics,* 78(4): 638-642, 1971.

Detection of reduction in branching enzyme activity in skin biopsies from children with type IV glycogen storage diseases (amylopectinosis), along with similar findings in both parents of 1 patient, suggests the possibility of antenatal detection of this untreatable, uniformly fatal disease, since branching enzyme activity of cultured fibroblasts from normal amniotic fluid is comparable to that of normal skin fibroblasts. Assays of branching, debranching, and α-1,4-glucosidase enzyme activities on fibroblasts derived from punch skin biopsies of 1 male and 2 female patients, from the parents of the male S, from 16 independent lines of 10 control Ss, and from fibroblasts developed from normal amniotic fluid cells, showed virtual absence of branching activity in the fibroblasts derived from the 3 patients, substantiating the association of this disorder with a genetic deficiency of the enzyme. Branching enzyme activity in fibroblasts derived from the 2 parents was reduced, supporting an autosomal recessive inheritance. The comparability of activity of the branching enzyme derived from control Ss with that from amniotic fluid, along with the ability to distinguish heterozygotes from affected Ss, suggests the possibility of antenatal diagnosis and genetic counseling in these cases. (9 refs.) - *M. S. Fish.*

Johns Hopkins Hospital
Baltimore, Maryland 21205

2681 **SUZUKI, YOSHIYUKI; BERMAN, PETER H.; & SUZUKI, KUNIHIKO.** Detection of Tay-Sachs disease heterozygotes by assay of hexosaminidase A in serum and leukocytes. *Journal of Pediatrics,* 78(4): 643-647, 1971.

Assay of the activity of the enzyme hexosaminidase A by cellulose acetate electrophoresis in serum and leukocytes of parents of children with Tay-Sachs disease is applicable for the detection of heterozygous carriers of the disorder. Analysis of serum and leukocyte suspensions obtained from 22 samples of serum of 6 parents of Tay-Sachs disease children and serum (22 Ss) and leukocytes (24 Ss) from normal controls and from the patients showed the complete absence of hexosaminidase A activity in the serum of 1 patient and a narrow range of activity in the serum of parents of the patients, with the level approximately ½ that of parents of control Ss. No overlap between the 2 groups occurred, and the difference was highly significant statistically. Analysis of the enzyme activity in leukocytes indicated that deficiency occurred for 3 patients. Activity in leukocytes of the parents of patients was much closer to the normal control range than activity in the serum. Serum appears to be preferable to leukocytes for the detection of the heterozygous carrier state, is far more easily prepared, and provides more consistent data. This identification of homozygous and heterozygous gene carriers of the disease may be made possible at an early (intrauterine and neonatal) stage of development by the use of amniotic fluid or cord blood and should be useful for genetic counseling. (14 refs.) - *M. S. Fish.*

University of Pennsylvania School of Medicine
Philadelphia, Pennsylvania 19104

2682 **WILROY, R. S., JR.; & ETTELDORF, JAMES N.** Familial hyperthyroidism including two siblings with neonatal Graves' disease. *Journal of Pediatrics,* 78(4): 625-632, 1971.

Neonatal hyperthyroidism, usually of brief duration, is reported for two siblings of a family in which 4 members of 3 generations suffered from Graves' disease. The Ss, Negro male and female sibs born a year apart whose mother had Graves' disease, had persistent neonatal thyrotoxicosis. Bone ages of both advanced rapidly and both developed craniosynostosis. Although antithyroid medication was given for several years, long-acting thyroid stimulator assays were elevated at ages 8 and 9 years, respectively, but were normal 7 years later. The female was more seriously affected early; her bone age was 5 years prior to age of 1 year, and cranial asymmetry and craniosynostosis were more profound, as were osseous abnormalities of the hands. Both were MR; however, each received specialized training, and the sister ultimately had the greater intellectual attainment. The father was reported to be normal; however, the maternal grandmother died at age 26 during surgical removal of a toxic goiter. The mother had 2 additional pregnancies. One resulted in the birth of a premature female who died at 6 days, the other produced a goitrous stillborn male. Neither of these 2 infants were studied. (14 refs.) - *M. S. Fish.*

University of Tennessee College of Medicine
Memphis, Tennessee 38103

2683 **ONISAWA, JINICHI; LEE, TING-YANG; NAKAMURA, NORIMASA; & OTSUKI, CHIEKO.** Chondroitin 4- and 6-sulfaturia in Morquio-Ullrich's syndrome. *Biochemical Medicine*, 5(1): 37-47, 1971.

Analytical studies of urinary acid mucopolysaccharides (AMPS) excreted by a patient with clinical symptoms similar to those found in Morquio-Ullrich's syndrome indicated that serine was the predominant amino acid bound in a small peptide attached to the chondroitin sulfate (CHS) chain and that the polysaccharide had a hybrid structure in its uronic acid composition. The patient, a product of a second-cousin marriage, was MR, dwarfed, and had bone changes characteristic of Morquio's disease. Analyses of urine collected from the S, from a group of control Ss, and from 4 cases of Hunter's syndrome showed that total AMPS excretion of the patient was 5 times greater than normal levels. Increased quantities of chondroitin-4- and chondroitin-6-sulfates were also excreted. Amino acid analysis of the peptide core of these sulfates showed that total amino acid content was below the normal values. The pattern of a smaller peptide core with a predominance of serine is similar to the composition of dermatan sulfate-peptides isolated from Hurler's urine. This finding, together with that of a hybrid structure in uronic acid composition, suggests that the primary defect in a number of genetic mucopolysaccharidoses may be related to both the formation of the peptide core and the AMPS chain. (30 refs.) - *M. S. Fish.*

University of Tokyo 3-1
Tokyo, Japan

2684 **SPERLING, O.; EILAM, G.; SCHMIDT, R.; MUNDEL, G.; & DE VRIES, A.** Purine base incorporation into erythrocyte nucleotides in hypoxanthine-guanine phosphoribosyltransferase deficiency. *Biochemical Medicine*, 5(2): 173-176, 1971.

Measurements of the specific activity of hypoxanthine-guanine phosphoribosyltransferase (HGPRT) in erythrocyte hemolysate and the rate of hypoxanthine and guanine incorporation into erythrocyte nucleotides in normal Ss, two patients with partial HGPRT deficiency, and a child with almost complete HGPRT deficiency is reported. Results showed that in erythrocytes devoid of HGPRT, incorporation of hypoxanthine or guanine into erythrocyte nucleotides does not occur, indicating the absence of an anabolic pathway involving nucleoside phosphorylase and kinase. This conclusion was reported previously from an examination of fibroblasts of patients with Lesch-Nyhan syndrome. In erythrocytes having partial HGPRT deficiency (specific activity of 0.45 to 2.4% compared to normal males) the finding that purine base incorporation occurs to the extent of 20.2 to 23.6% of normal into the total nucleotides indicates that enzyme assay in hemolysates as an index of impairment of cellular nucleotide synthesis is an unsatisfactory measure. The absence of neurological disturbances in patients with partial HGPRT deficiency might be due to the fact that, as observed in HGPRT-deficient erythrocytes, substantial incorporation of hypoxanthine and guanine into brain cell nucleotides might occur. (11 refs.) - *M. S. Fish.*

Rogoff-Wellcome Medical Research Institute
Petah Tikva, Israel

2685 **SOLLEE, NATALIE D.; & KANG, ELLEN S.** Treatment in phenylketonuria: Statistical questions. *Pediatrics*, 47(6): 1090-1091, 1971. (Letter)

The response to a letter raising questions about apparent inconsistencies in a study on treatment of phenylketonuria (PKU) states that the 2 sibling control groups utilized in the study do differ, not in mean IQ's, but in variability. Separate comparisons with sibling groups would have indicated only the differences reported. The authors also point out weaknesses in the use of analysis of variance for studies of this type, noting that the F ratio is only an approximate test of significance. They did not report the "strongly significant correlation between length of dietary treatment and IQ change for late-treated patients," since lack of IQ progress was the major criterion for terminating treatment. The original article stated the reasons for lack of inclusion of 9 of 19 Ss in the late-treated group. (2 refs.) - *M. S. Fish.*

300 Longwood Avenue
Boston, Massachusetts 02115

2686 FORD, ROBIN C. Treatment in phenylketonuria: Statistical questions. *Pediatrics,* 47(6): 1090, 1971. (Letter)

Questions are raised regarding the statistical evaluation in a recent study which reviews treatment of 82 classical and atypical phenylketonuric (PKU) children. While the study concluded that late-treated classical PKU Ss function at a slightly lower intellectual level than do their non-PKU sibs, and that early-treated PKU Ss do not differ from their sibling controls, the fact that no significant differences appear in the IQ scores of early- and late-treated Ss may have arisen either because the 2 sibling control groups may differ significantly from each other or because the 3 groups do not differ. The suggestion is made that appropriate planning for multiple comparisons of this type should have begun with a test for possible differences, e. g., through the use of analysis of variance, providing an F ratio. The lack of reporting of the significant correlation between the length of dietary treatment and the change in IQ and the omission of 9 Ss from this latter analysis are also criticized. (4 refs.) - *M. S. Fish.*

2300 Children's Plaza
Chicago, Illinois 60614

2687 ROE, THOMAS F.; HALLATT, JACK G.; DONNELL, GEORGE N.; & NG, WON G. Childbearing by a galactosemic woman. *Journal of Pediatrics,* 78(6): 1026-1030, 1971.

A recent study has, for the first time, demonstrated that a woman homozygous for galactosemia can produce a clinically normal infant. The S was a negro female first referred at 4 months of age for growth failure and persistent proteinuria. Galactosemia was suggested by the presence of hepatomegaly, of fine lenticular cataracts, and of a non-glucose-reducing urinary substance. When an oral glucose tolerance test strengthened the diagnosis, the S was placed and maintained on a lactose-restricted diet which improved her condition. At 19 yrs of age the S had an uncomplicated pregnancy. The father was found to have normal galactose-1-phosphate uridyl transferase activity. Management during pregnancy was normal except for the continued restriction of dietary galactose. Monitoring of maternal blood, urine, amniotic fluid, colostrum, and cord blood indicated that the influence of endogenous galactose production was minimal. (23 refs.) - *M. S. Fish.*

4650 Sunset Boulevard
Los Angeles, California 90054

2688 WINICK, MYRON. Cellular growth during early malnutrition. *Pediatrics,* 47(6): 969-978, 1971.

Available data indicate that postnatal undernutrition during the period of proliferative brain cell growth will retard the rate of cell division and result in a permanent reduction in the number of brain cells. In the rat prenatal malnutrition acts in a similar manner, and both pre- and post-natal undernutrition act synergistically, producing a profound reduction in brain cell number. Studies of normal and retarded growth in rats by serially measuring nucleic acid and protein content of various tissues have shown that in all nonregenerative organs, including the brain, cell division stops before the organ attains its final size. The time of cessation of DNA synthesis varies among different species of animals but occurs at or shortly after birth, whereas net protein synthesis continues throughout the growing period. Thus early and late growth differ fundamentally and explain observed differences in recovery patterns. Interference with cell division causes permanent effects on the brain, and malnutrition affects specific regions of the brain differently. Malnutrition causes greater retardation in regions having the more rapid rates of cell division. Two enzymes, DNA polymerase and RNAase, and the effects of malnutrition on DNA polymerase and RNAase, which appear to be involved in the rate of cell division, are under investigation, as are other chemical parameters of brain growth. Early postnatal malnutrition affects the human brain in a manner similar to that of the rat. While data on human prenatal malnutrition are limited, those obtained by examination of the placenta suggest retardation of placental cell growth due to maternal malnutrition. Indirect evidence indicates that a large reduction in fetal brain cell number may be due either to *in utero* malnourishment or to the effects of prematurity, making infants susceptible to subsequent undernutrition. (34 refs.) - *M. S. Fish.*

Cornell University Medical College
New York, New York 10021

2689 **EASTON, JOANNE; MARTIN, J. K.; & CAMPBELL, D. J.** More data on sex ratio in hyperphenylalaninemia. *Journal of Pediatrics,* 78(6): 1078, 1971. (Letter)

Additional data affecting the controversy over the presence or absence of male predominance in newborn hyperphenylalaninemia have been reported. Of 30,141 babies born over a 21-month period in Alberta, Canada, for whom serum phenylalanine was measured by automated quantitative fluorometry, 179 (104 males and 75 females) had initial levels > 3.5 mg/100 ml. Ninety (50 males and 40 females) had initial levels > 4.0 mg/100 ml. (2 refs.) - *M. S. Fish.*

University of Alberta Hospital
Edmonton, Alberta, Canada

2690 **KOCH, RICHARD; DOBSON, JAMES; HSIA, DAVID Y. Y.; & WOOLF, LOUIS I.** Conference on sex ratio in phenylketonuria. *Journal of Pediatrics,* 78(1): 157-159, 1971.

A recent conference on the sex ratio in phenylketonuria (PKU) has concluded that present screening programs are adequate, are not missing a significant number of PKU children, and should be continued. The conference was convened to consider the significance of a report of a Collaborative Study of Children Treated for PKU in which a sex ratio of 60 males to 30 females was observed. These findings brought up the question of whether present tests miss a significant number of PKU females. The conference concluded that if this observation of a disproportionate sex ratio had not occurred by chance it could be the result of criteria for inclusion in the study or of a slight variation in the influence of sex on phenylalanine metabolism. This latter factor could account either for inclusion of some males who do not have typical PKU or the exclusion of some females who do, or both. In cases of MR or other neurologic disorders, physicians should be aware of the possibility of a missed PKU case. An additional recommendation was for the need to verify the accuracy of the initial PKU diagnosis by challenging patients with normal phenylalanine intake at 6-month intervals. The conference also concluded that some children are being unnecessarily maintained on low phenylalanine diets, but that all children with persistent markedly elevated phenylalanine levels should be treated. Diagnosis and management of these cases are best carried out by a multidisciplinary group of experts knowledgeable about the disorder. (6 refs.) - *M. S. Fish.*

Children's Hospital of Los Angeles
Los Angeles, California

2691 **LUBCHENCO, LULA O.; & BARD, HARRY.** Incidence of hypoglycemia in newborn infants classified by birth weight and gestational age. *Pediatrics,* 47(5): 831-838, 1971.

A high incidence of neonatal hypoglycemia a few hours after birth results from a combination of reduced energy reserves in newborn infants with intrauterine growth retardation (IUGR) and the increased utilization of carbohydrates during birth hypoxia. From a group of 374 randomly selected infants, classified over a 2-year period into 9 categories by birth weight and gestational age, a clinical impression was obtained of the state of nutrition, the weight/length ratio, and glucose levels of umbilical cord samples before the first feeding at 3 to 6 hrs after birth. When hypoglycemia was determined in terms of levels <30 mg of serum glucose/100 ml, it occurred in 67, 25, 18, and 10% of the cases of preterm, small for gestational age (SGA); term SGA; postterm SGA; and full-term, appropriate for gestational age infants, respectively. A 35% incidence of hypoglycemia occurred for infants of diabetic mothers (IDM). These latter infants were generally large for gestational age and were delivered preterm. Except for IDM's, the physical examination and weight/length ratios confirmed IUGR in hypoglycemic infants. (22 refs.) - *M. S. Fish.*

Box 2776
4200 East Ninth Avenue
Denver, Colorado 80220

2692 **CLARKE, JOE T. R.; KNAACK, JUERGEN; CRAWHALL, JOHN C.; & *WOLFE, LEONHARD S.** Ceramide trihexosidosis (Fabry's disease) without skin lesions. *New England Journal of Medicine,* 284(5): 233-235, 1971.

The incidence of 2 unrelated cases of ceramide trihexosidosis (Fabry's disease) without the typical clinical cutaneous manifestations is described.

The Ss, 39- and 48-year-old males with unexplained proteinuria, had renal lesions and typical histochemical features of Fabry's disease (increased excretion of trihexosylaramide and markedly decreased activity of α-galactosidase), although neither had other manifestations of the disorder, which is usually characterized by angiokeratoma and corneal opacities. Renal biopsy provided the data related to the pattern of glycolipid excretion which has been established for the disease. Examination of leukocytes from the patients indicated a markedly decreased activity of α-galactosidase activity, suggesting the possibility that these patients may differ genetically from those with classical Fabry's disease. The appearance of these 2 unrelated cases within a few months at the same hospital suggests the possibility that the particular condition may be more common than has been previously reported. (14 refs.) - *M. S. Fish.*

*Montreal Neurological Institute
Montreal 112, Canada

2693 **WILSON, MIRIAM G.; WU, PAUL Y. K.; & WARE, ARNOLD C.** Sex ratio in hyperphenylalaninemia of newborn infants. *Journal of Pediatrics,* 78(1): 117-119, 1971.

A study has indicated that there is no significant sex difference in the occurrence of transient hyperphenylalaninemia in newborn (usually premature) infants. The experimental group was 57 infants of a total of 52,000 screened, with serum phenylalanine levels >/=4 mg%. Of these, 2 were later diagnosed as having atypical or variant phenylketonuria. The remaining 55 infants (46 of low birth weight) had an approximately equal sex distribution (26 male and 29 female). Sex distribution of low birth weight was also nearly equal. Of 49 infants having weekly phenylalanine tests from birth, no sex differences were observed in the rapidity with which serum phenylalanine levels reached >/=4 mg%. Approximately one-third reached this level in the first week; the remainder reached the level in the second week. When 34 surviving infants were followed to determine how long high serum phenylalanine levels were maintained, no significant sex differences in this measure were observed. The highest observed levels of serum phenylalanine were also comparable for both sexes of all 55 Ss. (8 refs.) - *M. S. Fish.*

University of Southern California
School of Medicine
Los Angeles, California 90033

2694 **SELMANOWITZ, VICTOR J.; VAN VOOLEN, G. ARTHUR; & STEIER, WILLIAM.** Fanconi's anemia and dyskeratosis congenita. *Journal of the American Medical Association,* 216(12): 2015, 1971. (Letter)

Dyskeratosis congenita (Zinsser-Engman-Cole syndrome) shares numerous features with Fanconi's anemia. These include progressive refractory pancytopenia, cutaneous dyschromia, obliteration of puncta lacrimalia, atretic external auditory canals, MR, overall frailty, hypogenitalism, and growth retardation. Dyskeratosis congenita is also associated with strong propensities for development of squamous cell carcinomas of mucosae and mucocutaneous junctions. In view of a recent report of the observation of squamous cell carcinomas of the oral mucosa, esophagus, anus, and vulva in adults with Fanconi's anemia, a question arises regarding a possible relationship between the two syndromes, despite the fact that each disorder also has a number of clinical features not noted in the other. The two disorders may be causally related, or the development of squamous cell carcinomas in adults with Fanconi's anemia may mark a transition between that disorder and dyskeratosis congenita. So far the karyotypic abnormalities of Fanconi's anemia have not been recorded for dyskeratosis congenita, however, whether the same inherited metabolic abnormality is present in all Fanconi's anemia cases is not known at present. (12 refs.) - *M. S. Fish.*

No address

2695 **WALKER, W. ALLAN; ULSTROM, ROBERT A.; & LOWMAN, JAMES T.** Albumin synthesis rates in patients with hypoproteinemia. *Journal of Pediatrics,* 78(5): 812-820, 1971.

A study of albumin synthesis rates of patients with hypoproteinemia has shown that excessive protein loss is apparently due to inadequate compensation for protein loss rather than to a primary defect in

protein synthesis. A group of 11 patients was divided into 3 groups: 4 Group I Ss (control) with normal serum albumin levels and no known abnormality in albumin metabolism; 4 Group II Ss having hyperproteinemia and with documented excessive loss of albumin via the bowel lumen or kidneys; and 3 Group III Ss with hypoproteinemia and with severe intrinsic liver disease. Injection of ^{75}selenomethionine, a radioactive amino acid precursor of albumin, followed by analysis of serial blood samples collected over a 5-day period (isolation of albumin by Sephadex gel filtration followed by determination of radioactivity) showed that for the Group II Ss the rate of incorporation by albumin of ^{75}selenomethionine (K_{inc}) was 9.38 x 10^5 cpm/liter of plasma/hour, compared to corresponding values of 4.88 x 10^5 and 1.58 x 10^5 for Group I controls and the Group III Ss with liver disease, respectively. Degradation curves also differed for the 3 groups: Group II had a slower early phase falloff compared to Group I, suggesting either a contracted albumin pool or slower equilibration. After 72 hours the degradation component of the curve was most and least steep in Group II and III Ss, respectively. The study shows that excessive loss of protein in the hypoproteinemic state is partially but not completely compensated for by increased albumin production. (32 refs.) - *M. S. Fish.*

Massachusetts General Hospital
Boston, Massachusetts 02114

2696 Hyperammonemia: Neonatal killer. First lethal case documented—how many go unrecognized? *Medical World News*, 12(22): 15-16, 1971.

Hyperammonemia may be a frequently unrecognized cause of neonatal deaths now attributed to sepsis. This is particularly true in cases of a newborn with coma and where there is a family history of neonatal death. A recent case of neonatal death of a male infant in which a similar fatality had occurred earlier for a male in the same family prompted an investigation of a possible causative metabolic error. Assay of liver enzymes at autopsy showed a complete lack of ornithine transcarbamylase (OTC), an enzyme in the urea cycle, with the result that the infant was unable to detoxify the ammonia which was the end product of protein and amino acid metabolism. Concentration of blood ammonia rose to 1,208 μg% compared to the normal level of 150 μg%. Although high blood ammonia has not been associated previously with newborn death, it has been identified as a cause of infantile somatic retardation and MR. The error may be sex-linked, transmitted by a gene on the X-chromosome, since previously reported cases of OTC deficiency occurred in females and were characterized by partial rather than complete enzyme absence. MR also occurred in these cases. Therapeutic approaches may involve restriction of dietary intake (probably effective for females); should the enzyme be found to be late-maturing rather than completely absent in the male Ss, cleansing of toxic accumulations of ammonia or other approaches may be helpful. Hyperammonemia has also been found associated with the deficiency of another urea cycle enzyme, carbamyl phosphate synthetase. In this case the female S also had a sister who died following a similar clinical course. (No refs.) - *M. S. Fish.*

2697 **BLASS, J. P.; LONSDALE, D.; UHLENDORF, B. W.; & HOM, E.** Intermittent ataxia with pyruvate-decarboxylase deficiency. *Lancet*, 1(7712): 1302, 1971.

Subsequent to a recent report of a defect in the enzyme pyruvate decarboxylase in the cultured fibroblasts of 1 of 2 unrelated patients with intermittent ataxia associated with hyperalaninuria and hyperpyruvicemia, a similar defect has been found in the other patient and in his affected sibling. Levels of the enzyme in sonicated cells were 9.3 and 9.7 nmole/g protein/minute in the 2 brothers, respectively, or less than one-third of the level found in normal controls. Excess thiamine pyrophosphate did not increase the activity of the enzyme, and no evidence for an inhibitor was found. Oxidation of 1-(^{14}C)-pyruvic acid was depressed, compared to a finding of normal oxidation of palmitic and glutamic acids. Binding of pyruvic acid was quantitatively similar in both the patients and controls. Pyruvate decarboxylase deficiency appears to represent another defect associated with intermittent ataxia, which also has been reported in Hartnup's disease and in intermittent branched-chain ketonaciduria (late onset of maple syrup urine disease). (3 refs.) - *M. S. Fish.*

University of California
Los Angeles, California

2698 **GROTH, C. G.; DREBORG, S.; OCKERMAN, P. A.; SVENNERHOLM, L.;**

HAGENFELDT, L.; LOFSTROM, B.; SAMUELSSON, K.; WERNER, B.; & WESTBERG, G. Splenic transplantation in a case of Gaucher's disease. *Lancet*, 1(7712): 1260-1264, 1971.

The results of providing a splenic whole organ homograft from a nonrelated donor to a patient with end-stage juvenile Gaucher's disease indicated that, before its rejection, the graft was the likely source of the favorable metabolic changes noted. The S, a 24-year-old male, had pancytopenia and hepatosplenomegaly diagnosed in infancy and a splenectomy at 2 years of age. The spleen tissue contained typical Gaucher's cells. Subsequent symptoms were kyphosis, rarefaction of the long bones, MR, psychomotor epilepsy at 12, and progressive grand mal at 15 years. After pretreatment with antilymphocyte globulin (ALG) and immunosuppressive drugs the S was given a spleen transplant, followed by spleen scans and analysis for total lipid hexose, glucocerebroside, ceramide, and plasma levels of lysosomal enzymes. After 2 days the patient's condition improved rapidly, and a significant drop in acid phosphatase and glucosaminidase occurred, along with a slight decrease in the number of Gaucher's cells in the bone marrow. Severe local reactions after 4 weeks required discontinuance of ALG, and the patient's condition deteriorated at 7 weeks. At 3 months postoperative severe skeletal pain and mental confusion developed, and, after contracting pneumonia, the patient died 90 days postoperatively. Spleen scans of the graft indicated that it functioned for about 40 days and then failed due to rejection. (30 refs.) - *M. S. Fish.*

University of Colorado Medical Center
Denver, Colorado 80220

2699 PATTON, VICTORIA M.; & *DEKABAN, ANATOLE S. G_{M1}-Gangliosidosis and juvenile cerebral lipidosis. Clinical, histochemical, and chemical study. *Archives of Neurology*, 24(6): 529-537, 1971.

A comparison of 2 patients with G_{M1}-gangliosidosis with a patient having juvenile cerebral lipidosis has shown that principal qualitative histological and histochemical findings are similar; however, in the former disorder the degree of distension of neurons with lipid staining material was much greater. Clinical findings in 2 cases of G_{M1}-gangliosidosis (a 4-year-old female and her 2-year-old brother) showed development of an early and profound spasticity but with normal-appearing optic fundi and little or no epileptic activity; whereas the patient with juvenile cerebral lipidosis (a 10-year-old female) had normal muscle tone, marked epileptic manifestations, and retinal deterioration along with total blindness. Histochemical analysis showed that in the G_{M1}-gangliosidosis the G_{M1}-ganglioside was 7 times normal, the asialo derivative was increased, and β-galactosidase activity was 5% of normal. This latter activity in a parent was in the normal range. In the other disorder cerebral cortex cholesterol and phospholipids were subnormal, and the ganglioside was normal. The gangliosides in the white matter were increased. (32 refs.) - *M. S. Fish.*

*National Institute of Neurological
Diseases and Stroke
Bethesda, Maryland 20014

2700 OLDENDORF, WILLIAM H.; SISSON, WARDEN B.; & SILVERSTEIN, ARTHUR. Brain uptake of selenomethionine SE 75. II. Reduced brain uptake of selenomethionine Se 75 in phenylketonuria. *Archives of Neurology*, 24(6): 524-528, 1971.

Brain uptake of selenomethionine Se 75 in Ss with phenylketonuria (PKU) is significantly reduced relative to uptake in a group of matched MR controls. Of 21 inst patients, 9 had PKU, and 12 without the disorder served as controls. The groups were matched for age, body weight, and the degree of retardation. A 2.5-microcurie dose of selenomethionine Se 75, dissolved in physiological saline, was injected intraveneously, and brain content following injection was monitored continuously for 20 min with a gamma radiation detector sampling a fixed volume of brain. In the PKU group uptake of the material was 0.72 ± 0.13 (dose counts per min/kg patient), compared to 1.12 ± 0.26 for the control group ($p<0.001$). The results are interpreted as being compatible with saturation of the blood brain barrier carrier system for selenomethionine Se 75 by high blood phenylalanine in the PKU Ss. The investigation may be useful in the elucidation of the mechanism of phenylalanine toxicity in PKU in that it shows that a defect may exist in the uptake of amino acids by the brain. (11 refs.) - *M. S. Fish.*

Wadsworth General Hospital
Los Angeles, California 90073

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2701 **EVANS, P. R.** Sotos' syndrome (cerebral gigantism) with peripheral dysostosis. *Archives of Disease in Childhood*, 46(246): 199-202, 1971.

An 18-year-old girl shows many features of Sotos' syndrome (cerebral gigantism) except that she has short, stumpy hands and feet (peripheral dysostosis). At age 3 she was in the 97th percentile for height, but growth stopped at age 14, and she is now in the 10th percentile. She is sexually mature. She did demonstrate delayed psychomotor development, macrocrania, clumsiness, and abnormal EEG. Despite mild MR she appears reasonably well adjusted. (3 refs.) - *E. L. Rowan.*

Guy's Hospital
London, S.E. 1, England

2702 **KELLY, SALLY.** Septicemia in galactosemia. *Journal of the American Medical Association*, 216(2): 330, 1971. (Letter)

Clinical signs of galactosemia and concomitant gram negative septicemia have been presented by at least 7 infants. The concurrence of galactosemia and gram negative septicemia in these infants may be a coincidence or may have a biochemical etiology. The infant with galactosemia who vomits excessively or has diarrhea may be especially susceptible to septicemia. The source of infection may be the urinary tract. (8 refs.) - *J. K. Wyatt.*

2703 **CRUMP, IAN A.; & DANKS, DAVID M.** Simple method for cutting transverse sections of hair. Comments on shape of hair in Hurler and Sanfilippo syndromes. *Archives of Disease in Childhood*, 46(247): 383-386, 1971.

Triangular hairs were present in samples of hair from 10 of 12 mucopolysaccharidoses patients, 3 of 10 normal individuals, and 6 of 11 patients whose hair felt coarse. The 2 exceptions among mucopolysaccharidoses patients were young children; later samples of their hair showed the presence of triangular hairs. Although this finding may not have diagnostic value in regard to Hurler's or Sanfilippo's syndromes, if triangular hairs prove to be a universal feature of Sanfilippo syndrome, their absence may help in exclusion of the diagnosis for MR children with coarse features. (1 ref.) - *J. K. Wyatt.*

University of Melbourne
Melbourne, Australia

2704 **MALMQUIST, JORGEN; JAGENBURG, RUDOLPH; & LINDSTEDT, GORAN.** Familial protein intolerance. Possible nature of enzyme defect. *New England Journal of Medicine*, 284(18): 997-1002, 1971.

A female patient (CA 36 yrs) displayed the clinical and biochemical features of familial protein intolerance, a hereditary defect of ammonium ion metabolism. However, familial occurrence was not documented in this case. The patient was MR (IQ 70), had x-ray evidence of brain atrophy, and displayed marked skeletal fragility. Intolerance of foods containing protein was moderate. She had a moderately elevated postprandial blood ammonia concentration. Intravenous administration of L-alanine resulted in marked ammonia elevation and marked hyperglycemia. A defect was present in the mechanisms that transfer amino nitrogen to the urea-synthesizing system. This is the thirteenth case of familial protein intolerance identified in Finland. (33 refs.) - *J. K. Wyatt.*

University of LUND
S-214 01 Malmo, Sweden

2705 **HABBICK, B. F.; McNEISH, A. S.; & STEPHENSON, J. B. P.** Diagnosis of ketotic hypoglycaemia of childhood. *Archives of Disease in Childhood*, 46(247): 295-300, 1971.

Follow-up studies on 20 children (14 boys, 6 girls) with a diagnosis of ketotic hypoglycemia during the previous 8 years revealed that the children were 'dysmature' at birth and remained small and thin. Two were MR, and 4 were the smaller of nonidentical twins. Age at diagnosis ranged from 13 months to 5 years 11 months. The majority had acute early morning convulsions after the age of 1 year accompanied by drowsiness or unconsciousness and marked ketonuria. Symptoms had been successfully controlled by treatment with carbohydrate supplement at bedtime in 16 of 19 cases. Failure to control symptoms seemed due to

lack of parent cooperation in 1 case and to erroneous diagnosis in 2 cases. (11 refs.) - *J. K. Wyatt.*

2706 LE DUNE, MARTHE A. Insulin studies in temporary neonatal hyperglycaemia. *Archives of Disease in Childhood,* 46(247): 392-394, 1971.

During the course of an illness an infant girl with temporary neonatal hyperglycemia had very high levels of immuno-reactive insulin. Tetany occurred on the eighth day of life. On the eleventh day she evidenced severe acidosis, and on the thirteenth day she had marked glycosuria. A glucose tolerance test on the thirteenth day revealed a striking rise in insulin levels as a result of the oral dose of glucose, suggesting that the immuno-reactive insulin in the infant's plasma was functionally inactive. Exogenous insulin was used to control the diabetic condition. This was administered for 3 days. By the seventeenth day of life the urine was free of reducing substances and the fasting true blood glucose was 57 mg/100 ml. A transfusion of whole blood (75 ml) was given at this time and her condition continued to improve. Insulin studies at 1 month showed a normal configuration of blood glucose levels and a fasting insulin level which was within the normal range but seemed exaggerated in terms of response to insulin. At 4 months, intravenous tolbutamide and glucose tolerance tests were within normal limits. The child is now microcephalic and has spastic tetraplegia. High levels of immunoreactive insulin in the presence of obvious hyperglycemia may be due to insulin production which is biologically inactive. (8 refs.) - *J. K. Wyatt.*

Stobhill General Hospital
Glasgow N.1, Scotland

2707 MILNER, R.D.G. Metabolic and hormonal responses to oral amino acids in infantile malnutrition. *Archives of Disease in Childhood,* 46(247): 301-305, 1971.

Eight children with infantile malnutrition and 7 children who had clinically recovered from malnutrition were given a mixture of 10 essential amino acids after admission to hospital. Blood samples were obtained after an overnight fast and at different times after administration of the amino acid load. They were analyzed for glucose, free fatty acid, α-amino nitrogen, insulin, and growth hormone. The amino acid load caused a rise in plasma α-amino nitrogren and glucose and a fall in growth hormone levels in all children. There were no changes in the levels of plasma free fatty acids or insulin. (20 refs.) - *J. K. Wyatt.*

St. Mary's Hospital
Manchester 13, England

2708 RAITI, S.; & NEWNS, G. H. Linear growth in treated congenital adrenal hyperplasia. *Archives of Disease in Childhood,* 46(247): 376-378, 1971.

Twenty-seven of 35 cases with the salt-losing type of congenital adrenal hyperplasia and 10 cases with the simple virilizing type grew within the third to the ninety-seventh centiles for height. Among the salt-losers, 8 grew below the third centile; all of these had been overtreated with cortisone (in excess of 36 mg/m^2/24 hrs.) and had delayed bone ages. Four of these 8 patients are now receiving oral doses of cortisone which are compatible with expected requirements, and 2 have already shown catch-up growth. Total daily cortisone replacement doses should not exceed twice the expected cortisol production rate for body size. A minimum cortisone dose of 15 mg/day in divided doses is recommended during the first 2 years of life. Growth failure appears related to excessive cortisone therapy for the patient's needs. (8 refs.) - *J. K. Wyatt.*

National Pituitary Agency
Baltimore, Maryland 21201

2709 VAN ACKER, K. J.; VINCKE, H.; QUATACKER, J.; SENESAEL, L.; & VAN DEN BRANDE, J. Congenital oligonephronic renal hypoplasia with hypertrophy of nephrons (oligonephronia). *Archives of Disease in Childhood,* 46(247): 321-326, 1971.

Although time of onset and severity varied, the clinical picture in 4 cases of oligonephronia was one of progressive renal insufficiency. One patient presented a familial syndrome of multiple congenital anomalies which included psychomotor retardation; abnormalities of the outer ear, eye,

fingers, and skeleton; branchial fistulas; dwarfism; and otovestibular, neurological, and mental disturbances. The patient had 2 very small kidneys. A sister had a simple unilateral renal hypoplasia. Since associated congenital anomalies are rare in oligonephronia, the question arises as to whether this syndrome of multiple anomalies is coincidental or related to oligonephronia. (10 refs.) - *J. K. Wyatt.*

Kindergeneeskunde Ryksuniversiteit
Gent, Belgium

2710 **MIGEON, BARBARA RUBEN.** Studies of skin fibroblasts from 10 families with HGPRT deficiency, with reference to X-chromosomal inactivation. *American Journal of Human Genetics,* 23(2): 199-210, 1971.

Family studies of hypoxanthine-guanine phosphoribosyl transferase (HGPRT) deficiency, utilizing procedures which take advantage of the X-chromosomal inactivation, have resulted in the detection of a relatively large group of heterozygotes. The study population was 10 unrelated HGPRT-deficient males and 35 members of their kindred. Features of this syndrome include developmental retardation, spastic cerebral palsy, choreoathetosis, and self-mutilation. Fibroblast cultures obtained from skin biopsies were maintained in growth medium, uncloned cells were exposed to medium which selected either mutant or wild types, and early subculture cells were utilized for cloning experiments and for autoradiographic studies of incorporation of ^{3}H hypoxanthine. The observation of similar cloning efficiency for mutant and wild types indicated that *in vitro* selection against enzyme-deficient skin fibroblasts did not occur. Individual variation in the proportion of enzyme-deficient cells likely accounts for the observed phenotypic variability for carriers of the deficiency. In the total group of 4,215 clones enzyme-deficient and wild-type clones occurred with equal frequency. One male S and members of his kindred were found to have a unique enzyme phenotype. (21 refs.) - *M. S. Fish.*

Johns Hopkins University
School of Medicine
Baltimore, Maryland 21205

2711 **MENKES, JOHN H.; O'BRIEN, JOHN S.; OKADA, SHINTARO; GRIPPO, JORGE; ANDREWS, JOHN M.; & CANCILLA, PASQUALE A.** Juvenile G_{M2} gangliosidosis. Biochemical and ultrastructural studies on a new variant of Tay-Sachs disease. *Archives of Neurology,* 25(1): 14-22, 1971.

Findings on a patient with clinical symptoms of late infantile amaurotic idiocy (Tay-Sachs disease) have indicated that the patient had juvenile G_{M2} ganglioside storage disease characterized by only partially active hexosaminidase A. The S was a white, non-Jewish female seen at 3 9/12 years because of an impaired gait and seizures which anticonvulsive medicine did not control. Intellectual development was below normal for the the chronological age and deteriorated during the following year, along with loss of ability to stand or crawl. Analysis of brain biopsy material at 5½ years showed enlarged neurons in the cortex and ultrastructure findings characteristic of those reported for late-onset G_{M2} gangliosidosis. Neurons contained abnormal cytoplasmic inclusions of several types: membranous cytoplasmic bodies described for Tay-Sachs disease and G_{M1} gangliosidosis; zebra bodies described in gargoylism; inclusions typical of lipofuscin pigment; larger aggregates of the preceding types; and irregularly shaped amorphous bodies. Both gray and white matter had accumulation of G_{M2} (G_5) ganglioside; however, the increase was less than that found in Tay-Sachs disease although greater than that observed for other disorders. Hexosaminidase A activity in serum and skin fibroblasts was reduced to 6.6% and 12% of the total hexosaminidase activity, compared to 57.3% and 49.0% in controls and complete absence of activity in Tay-Sachs disease. This partial enzyme deficiency likely accounted for the later onset and the slower progression of CNS degeneration. (33 refs.) - *M. S. Fish.*

UCLA Center for the Health Sciences
Los Angeles, California 90024

2712 **LAMBOTTE, C.; ISRAEL, E.; & DURENNE, J.-M.** La deficience en glucose-6-phosphate deshydrogenase (Glucose-6-phosphate dehydrogenase deficiency). *Acta Paediatrica Belgica,* 25(2): 88-108, 1971.

Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency, a hereditary disorder transmitted by the X chromosome, affects hemizygous males and homozygous females. Favic individuals in the Mediterranean region have been found to manifest G-6-PD

deficiency, and it appears that the presence of favism indicates the existence of excessive tyrosinatic activity or an exceptionally slow degradation of dopaquinone as well. Evidence has confirmed that leukocytic G-6-PD deficiency is almost total, with phagocytic and bacteriocidal activity significantly affected. G-6-PD deficiency is 1 of the main causes of chronic nonspherocytic hemolytic anemia, neonatal icterus, and acute hemolysis. Systematic identification of this enzymatic defect is especially important in Belgium as a result of immigration from regions where the 2 primary biochemical types of the deficiency are prevalent (Mediterranean basin and Black Africa). (87 refs.) - *B. J. Grylack.*

Universite de Liege
B-4000 Liege, Belgium

2713 **ISRAEL, E.; LAMBOTTE, C.; & CHAPELLE, P.** Hemoglobinose S et deficience en glucose-6-phosphate deshydrogenase (Hemoglobinopathy S and glucose-6-phosphate dehydrogenase deficiency). *Acta Paediatrica Belgica*, 25(2): 109-118, 1971.

A eutrophic 2-month-old infant suffering from sickle cell anemia with an early onset and glucose-6-phosphate dehydrogenase (G-6-PD) deficiency presented with hemolytic anemia with anisocytosis, poikilocytosis, drepanocytes, and orthochromatophilic normoblasts. There was an absence of hemoglobin A_1, and systematic examination revealed a significant deficit in the erythrocytic enzyme. The infant had to be transfused at 3.5 mos and again at 5.5 mos, since the hemoglobin level decreased progressively to 6%. While the possibility that early hemolysis is attributable to sickle cell anemia alone cannot be excluded, the present case supports the unfavorable influence of G-6-PD deficiency on the prognosis for sickle cell anemia. (30 refs.) - *B. J. Grylack.*

Universite de Liege
B-4000 Liege, Belgium

2714 **SINGER, J. D.; & BROCK, D. J. H.** Half-normal adenylate kinase activity in three generations. *Annals of Human Genetics*, 35(1): 109-114, 1971.

A SMR patient with a half-normal specific activity of red-cell adenylate kinase (AK) was investigated. Cytologically, no sex chromatin bodies were detected in the nuclei of buccal epithelial cells or drumsticks in polymorphonuclear granulocytes. The karyotype showed 22 pairs of autosomes and normal male sex chromosomes, and there was extreme variation in length affecting the long arms of the number 16's in most cells. Biochemically, electrophoresis and enzyme kinetic study of hemolysates from the propositus showed a normal AK 1 phenotype. Segregation of the half-normal AK value was discovered in 3 generations of the family. The abnormal findings in this case may have been the consequence of a silent allele segregating through 3 generations of the family of the propositus. (14 refs.) - *B. J. Grylack.*

Harperbury Hospital Nr St. Albans
Hertfordshire, England

2715 **EYRE, CHRISTINE; DAVISON, A. N.; & SCOTT, J. T.** Phosphoribosyl transferase activity in normal subjects, gout patients, and children with mental retardation. *Journal of Neurology, Neurosurgery, and Psychiatry*, 34(1): 23-24, 1971.

The activity of hypoxanthine-guanine phosphoribosyl transferase (HGPRTase) is absent in the Lesch-Nyhan syndrome and partially deficient in affected members of families with gout; however, activity of this enzyme was found to be normal in 10 normal adults, 37 subjects with gout, and 21 MR children. A high uric acid to creatinine ratio in urine was not associated with HGPRTase deficiency. Such an enzyme deficiency must be rare in such cases of gout or MR as are usually encountered. (7 refs.) - *E. L. Rowan.*

Charing Cross Hospital
London, England

2716 **HASLAM, ROBERT H. A.; & CLARK, DAVID B.** Progressive cerebellar ataxia associated with Hand-Schuller-Christian disease. *Developmental Medicine and Child Neurology*, 13(9): 174-179, 1971.

A young man had a 10-year-history of progressive cerebellar ataxia and dysarthria before the appearance of diabetes insipidus and osteolytic lesions suggested a diagnosis of Hand-Schuller-Christian disease. Serum uric acid was found to be elevated, but the reason for this is

unknown. Because of the progressive nature of the neurological defect, a course of immunosuppressive agents followed by radiation therapy was given. Followup 10 months later showed stabilization of neurological function. (21 refs.) - *E. L. Rowan.*

John F. Kennedy Institute
Baltimore, Maryland 21205

2717 **SIBINGA, M. S.; FRIEDMAN, C. J.; STEISEL, I. M.; & BAKER, E. C.** The depressing effect of diet on physical growth in phenylketonuria. *Developmental Medicine and Child Neurology*, 13(1): 63-70, 1971.

Growth patterns of children with phenylketonuria (PKU), treated by a low phenylalanine diet, show that significant decrements in height and weight occur for these Ss, irrespective of the age when the diet was begun. The study population was 86 children, ages 7 months to 18 years (34 female and 52 male), with PKU. The Ss were divided into 3 groups: on diet (those treated continuously with a low phenylalanine diet), 60; off diet (those previously treated with the diet but for whom the diet had been discontinued), 12; and never on diet, 14. The weight at birth was available for 62 Ss and served as baseline data. Height and weight were recorded for all Ss during routine hospital visits. When the Kruskall-Wallis One-Way Analysis of Variance was employed to determine any relationship between diet status and height and weight percentile scores, PKU infants fell within normal limits of variability for weight and length at birth; severe adverse effects of the diet on growth were observed in comparison of the 3 groups; no significant relationship occurred between height and age or between weight and age at which diet was begun; and an inverse relationship between duration of diet therapy and height and weight percentile measurements occurred, irrespective of the age when diet was begun. Measurements of height and weight of diet-treated PKU children indicated a distribution differing from that of the normal population. (11 refs.) - *M. S. Fish.*

Temple University School of Medicine
Philadelphia, Pennsylvania 19133

2718 **MASTROPAOLO, C.; PAMPIGLIONE, G.; & STEPHENS, ROSEMARY.** EEG studies in 22 children with sulphatide lipidosis (metachromatic leucodystrophy). *Developmental Medicine and Child Neurology*, 13(1): 20-31, 1971.

EEG features of children with sulphatide lipidosis (SL; metachromatic leukodystrophy) differ substantially from those of children with other inborn errors of metabolism which affect the CNS, and EEG patterns may be useful in the early differential diagnosis of children with obscure neurological illnesses. On 22 children with SL, selected on the basis of histologically proven evidence of the disease (brain biopsy or autopsy or biopsy of rectal tissue, peripheral nerve, skin, or urine), a total of 55 EEG studies were carried out with a uniform technique (an 8-channel type D or T Offner apparatus). Concomitant neurological evaluations were conducted. The Ss were divided into groups based on the ages at which symptoms began: Group A, below 2½ years (13 Ss), and Group B, between 4 and 10 years (9 Ss). Evaluation of EEG studies showed a slightly greater amount of irregular fast activity for Group B compared to Group A Ss. In the 7 Ss with seizures (3 of Group A and 4 of Group B), EEG discharges tended to be irregular. Of the Group A children who had EEG's early in the stage of disease development, progressive increase in rhythmic slow components occurred, with activity more obvious over the posterior compared to the anterior half of the two hemispheres. In 2 of the older children of Group B, at risk because of an affected sibling, EEG studies begun before the onset of symptoms indicated definite abnormalities long before clinical signs were evident, suggesting that alteration in the electrogenetic mechanisms may be an early sign of SL. Among the 22 Ss spikes or complex wave forms were uncommon, in contrast to EEG features reported for comparable patients with other sphingolipidoses where a predominance of spikes occurs. (28 refs.) - *M. S. Fish.*

Hospital for Sick Children
London W. C. 1, England

2719 **SIBINGA, MAARTEN S.; & FRIEDMAN, C. JACK.** Complexities of parental understanding of phenylketonuria. *Pediatrics*, 48(2): 216-224, 1971.

A questionnaire was directed to 40 mothers and 39 fathers representing 42 families of children with phenylketonuria (PKU) in order to determine the extent of parental understanding of PKU and distortion tendencies and the relationship of these factors to parent education and intellectual and behavioral adjustment of the children. Only 19% of the 79 parents gave adequately correct answers. Parents with greater education did not respond significantly more accurately or completely to the questionnaire than parents with less schooling. The intellectual status of the child did not vary with either the mother's or father's understanding of the disease. The better the combined understanding ($p<.05$), the more intelligent the child was found to be. Approximately half of each parent group distorted its explanations of PKU. Fathers with greater distortion ratings had children with lower IQ scores. The results of the study suggest the need for a reexamination of certain facets of the management of the chronically ill child. (8 refs.) - *B. J. Grylack.*

St. Christopher's Hospital for Children
Philadelphia, Pennsylvania 19133

2720 **MOLLICA, FLORINDO; PAVONE, LORENZO; & ANTENER, ILSE.** Pure familial hyperprolinemia: Isolated inborn error of aminoacid metabolism without other anomalies in a Sicilian family. *Pediatrics*, 48(2): 225-231, 1971.

Type I hyperprolinemia was discovered in some members of a Sicilian family, in which there was no evidence of renal involvement or other congenital anomalies. The normal urinary excretion of Δ_1-pyrroline-5-carboxylic acid (PC) proved that the case belonged to type I of familial hyperprolinemia, despite increased serum proline levels. Based on the study of this family and another reported in 1970, this type of familial hyperprolinemia is presented as an example of an inborn error of aminoacid metabolism that is absolutely symptomless in all the members of the pedigree and can be defined as "pure familial hyperprolinemia." It is probably transmitted by an autosomal recessive gene, which may produce variable degrees of hyperprolinemia even in the heterozygotes. It is uncertain at present whether the hyperprolinemia of nephropathic families and that of the 2 families without nephropathy is due to the action of a unique gene. (26 refs.) - *B. J. Grylack.*

Clinica Pediatrica dell'Universita di Catania
95125 Catania, Italy

2721 **HUMBERT, JAMES R.; & *GOTLIN, RONALD W.** Growth hormone levels in normoglycemic and hypoglycemic infants born small for gestational age. *Pediatrics*, 48(2): 190-199, 1971.

Forty-six newborn infants, including 18 full-term small for gestational age (FSGA) infants, 18 full-term appropriate for gestational age (FAGA), and 10 premature (PR) infants, were used in a prospective study of serial basal growth hormone (GH) levels and simultaneous serum glucose determinations carried out to discover whether lack of GH in FSGA infants might contribute to their small size and hypoglycemic tendency. No significant difference was found between the GH values of FAGA, FSGA, and PR infants at any time during the first 3 days of life or between daily mean glucose values in the 3 groups. No significant correlation was observed between the simultaneously obtained GH and glucose values in the study (coefficient of correlation 0.27, N=89). Of 4 infants who were hypoglycemic, all attained normal glucose levels by the end of the first week of life. GH levels of these infants were neither abnormally elevated nor depressed at the time hypoglycemia was recorded. GH level appears to be a contributing factor in the small size or hypoglycemic tendency of FSGA newborn infants. (21 refs.) - *B. J. Grylack.*

*The University of Colorado Medical Center
Denver, Colorado 80220

2722 **KINT, J. A.; & DACREMONT, G.** ABO blood groups and lysosomal diseases. *New England Journal of Medicine*, 285(2): 121, 1971. (Letter)

Based upon the study of ABO blood groups and lysosomal diseases, it has been found that patients with Fabry's disease belonging to blood Group B have an extra amount of nonhydrolyzable molecules, which may lead to a more severe clinical picture than is noted in patients with other blood groups. Analysis of the oligosaccha-

rides in the urine of patients with Fabry's disease may reveal an important difference according to the blood group of the patient. In fucosidosis, patients with blood Group O have fewer nondegradable molecules due to a lack of blood-group-active macromolecules, as compared to patients with blood Group A, B, or AB. Researchers of these diseases should note any relation between clinical and biochemical abnormalities and the blood groups of patients. (3 refs.) - *B. J. Grylack.*

Kinderkliniek
Akademisch Ziekenhuis
Gent, Belgium

2723 **DAWSON, GLYN; & SPRANGER, JURGEN W.** Fucosidosis: A glycosphingolipidosis. *New England Journal of Medicine*, 285(2): 122, 1971. (Letter)

Examination of a small liver biopsy from a patient with fucosidosis allowed the glycosphingolipid accumulated due to absence of α-fucosidase activity to be characterized sufficiently to classify the disease as a glycosphingolipidosis. The concentration of the glycosphingolipid found in this case was 2.8 μmoles per g fresh weight, which is comparable to the levels of glucosylceramide in infantile Gaucher liver (11.4 μmoles per g fresh weight) and trihexosylceramide in Fabry liver. This amount is at least 100 times that found in normal liver. After methanolysis and gas-chromatographic analysis of the trimethylsilyl methyl glycosides, the glycosphingolipid in this case was found to contain fucose, galactose, glucose, N-acetylglucosamine and ceramide in a molar ratio of 1:2:1:1:1. Since many glycoproteins contain α-fucoside linkages, it is possible that their degradation is also affected in this disease. (7 refs.) - *B. J. Grylack.*

University of Chicago
Chicago, Illinois

2724 **FEIGIN, RALPH D.; SHACKELFORD, PENELOPE G.; DeVIVO, DARRYL D.; & HAYMOND, MOREY W.** Floxuridine treatment of congenital cytomegalic inclusion disease. *Pediatrics*, 48(2): 318-322, 1971.

Floxuridine (5-fluorouracil-2-deoxyriboside, or FUDR) was used in a neonate with severe cytomegalic inclusion disease. Treatment with FUDR was begun at 12 days of age and consisted of intravenous administration of 0.5 mg/kg daily as a single continuous 12-hour infusion every day for 5 consecutive days. Cultures for cytomegalovirus (CMV) continued to be positive throughout therapy with FUDR, and urine and salivary cultures were negative only after 28 days of age, with the possibility of persistent viremia being present. Based upon clinical and laboratory findings, a therapeutic effect of FUDR on congenital cytomegalic inclusion disease could not be demonstrated. The course of the patient after FUDR administration revealed no sign of toxicity. Although success could not be shown in this patient, more prolonged FUDR administration or treatment with other compounds such as idoxuridine or cytosine arabinoside should continue to be attempted. (11 refs.) - *B. J. Grylack.*

St. Louis Children's Hospital
St. Louis, Missouri 63110

2725 **MOSKOWITZ, MICHAEL A.; WINICKOFF, RICHARD N.; & HEINZ, E. RALPH.** Familial calcification of the basal ganglions. A metabolic and genetic study. *New England Journal of Medicine*, 285(2): 72-77, 1971.

A family with 5 members in a 2-generation span having familial bilaterally symmetric calcification of the basal ganglions was observed clinically and was studied metabolically and genetically. The affected members described a spectrum from entirely asymptomatic patients to those affected by severe extrapyramidal dyskinesias. Control studies for basal excretion of 3′, 5′-adenosine monophosphate (3′,5′-AMP) were performed on 4 affected family members, and 18 USP units of purified parathyroid hormone were administered to 3 of these patients. A greater than normal base-line output of 3′5′-AMP and greater than normal excretion of 3′,5′-AMP in response to parathyroid hormone were documented. The pattern of inheritance for this kindred is consistent with autosomal dominance. Male-to-male transmission is recorded, and sexes are affected almost equally. The syndrome of familial basal-ganglion calcification differs clinically, genetically, and biochemically from other disorders of calcium metabolism which lead to basal ganglion calcification. (36 refs.) - *B. J. Grylack.*

Yale University School of Medicine
New Haven, Connecticut 06510

2726 **LEVY, HARVEY Z.; SHIH, VIVIAN E.; KAROLKEWICZ, VALERIE; FRENCH, WILMA A.; CARR, JANE R.; CASS, VICTORIA; KENNEDY, JOSEPH L., JR.; & MacCREADY, ROBERT A.** Persistent mild hyperphenylalaninemia in the untreated state. A prospective study. *New England Journal of Medicine*, 285(8): 424-429, 1971.

Nine infants with persistent mild hyperphenylalaninemia, identified as a direct result of routine phenylketonuria (PKU) screening, were followed from the neonatal period into early childhood. The disorder was also detected in 1 mother and 3 siblings as a result of routine screening of members of families in which a neonate was affected by it. None of the 13 Ss had received any form of therapy. All but 1 of the children identified as a direct result of routine newborn screening manifested the disorder within the first 3 to 6 days of life. One infant had a normal phenylalanine concentration (less than 2 mg per 100 ml) at the age of 3 days but showed 6 mg per 100 ml at 4 weeks. Phenylalanine concentrations never exceeded 12 mg per 100 ml but were usually 6 mg per 100 ml or higher. General clinical evaluation during early infancy and at the time of psychometric testing did not reveal any abnormalities in any S, with growth and development always remaining within normal range. In some cases, the biochemically affected child manifested an IQ in the "superior" range, and the adult scored in the "bright-normal" range. Good school grades were reported for all affected children who were attending or had attended classes. Since the children have not suffered intellectual impairment even though they were not placed on treatment, a low-phenylalanine diet does not seem to be indicated for persistent mild hyperphenylalaninemia. (36 refs.) - *B. J. Grylack.*

Division of Diagnostic Laboratories
State Laboratory Institute
Boston, Massachusetts 02130

2727 Early malnutrition. *Medical World News*, 12(April 2): 5,1971.

An experiment performed by Richard J. Wurtman and William J. Shoemaker of MIT is reported. Two groups of pregnant rats were fed a 24% protein diet and 2 groups were fed an 8% protein diet. Rat pups were decapitated and their brains examined at 12 and 24 weeks. Some deprived rats' brains were three-quarters the weight of the well-nourished, but, paradoxically, the activity of tyrosine hydroxylase was significantly up in the deprived rats' brains. It is suggested, therefore, that malnutrition does not decrease the number of noradrenergic neurons. This is, however, only a preliminary finding. (No refs.) - *T. C. Turner.*

2728 **KALLEN, DAVID J.** Nutrition and society. *Journal of the American Medical Association*, 215(1): 94-100, 1971.

The best documented of all findings relating nutritional intake to human development is the reduction in growth rate with malnutrition. Severe malnutrition after birth, resulting in Kwashiorkor or marasmus at an early age, interferes with the normal development of the brain. Malnutrition is associated with apathy, listlessness, and unresponsiveness to external and internal stimuli. There is some evidence to indicate that marasmus and Kwashiorkor produce deficits in intellectual ability, and malnutrition has also been shown to be a definite deterrent to a good mother-child relationship. In addition to causing individual tragedy, malnutrition creates a disvalued and dysfunctioning group within society. (41 refs.) - B. J. Grylack.

Michigan State University
East Lansing, Michigan 48823

2729 **WHITE, PHILIP L.** The world stops for the hungry. *Journal of the American Medical Association*, 215(1): 110-111, 1971.

Severe malnutrition during the latter stages of pregnancy and the first few months of life will jeopardize physical and mental development. The dull or unresponsive infant or young child who is poor and malnourished may respond to an improved diet, but he will continue to have intellectual problems if his environment is culturally and economically devoid of stimuli. The frustration of attempting to isolate the role of malnutrition from other environmental factors in mental impairment has served to reemphasize the importance of stimulation for the development of the intellect. The complex determinants of

social interaction which define a disadvantaged segment of an otherwise affluent society must be understood if nutritional programs are to be utilized effectively. (No refs.) - *B. J. Grylack.*

AMA Council of Foods and Nutrition
Chicago, Illinois

2730 The hepatic glycogenoses. *Lancet*, 1(7712): 1339-1340, 1971.

The combination of hepatomegaly and neurological symptoms in a child often signifies the possibility of glycogen-storage disease. The glycogenoses can be detected before symptoms appear, and it should be possible to prevent neurological damage through early initiation of treatment. Even without enzyme assay and liver biopsy, much useful information can be obtained with simple biochemical tests. Long-term therapy of the glycogenoses is aimed at maintaining a normal blood-glucose in order to prevent possible neurological complications. All patients with suspected glycogen-storage disease need full investigation, and enzyme assays of a liver-biopsy specimen are indicated if fasting hypoglycemia and lack of response to glucagon are found in a child together with the appropriate clinical features. (8 refs.) - *B. J. Grylack.*

2731 **CROME, L.; & KANJILAL, G. C.** Cockayne's syndrome: Case report. *Journal of Neurology, Neurosurgery and Psychiatry*, 34(2): 171-178, 1971.

A nonfamilial case of Cockayne's syndrome was seen with the usual manifestations, including SMR, dwarfism with microcephaly, severe perceptive deafness, pigmented retinopathy, and progeria-like face. This female patient lived until the age of 22, when she died of status epilepticus due to grand mal epilepsy. Autopsy examination of the central nervous system showed deposition of globules of PAS-positive hyaline material within and upon the outer surface of capillaries. Many of the larger vessels, chiefly arterioles, were also abnormal, showing thickening and impregnation of their walls by the hyaline material, partly or fully calcified. As with all syndromes formed by multiple components, some variability of the manifestation of the Cockayne syndrome must be accepted. The relative frequency of familial cases of Cockayne's syndrome suggests a recessive mode of inheritance and the operation of some inherited metabolic defect. (15 refs.) - *B. J. Grylack.*

Queen Mary's Hospital for Children
Carshalton, Surrey, England

2732 **O'LEARY, JAY.** PKU counseling. *Hospital Practice*, 6(8): 13, 1972. (Letter)

A commentary on a letter requesting advice on genetic counseling for phenylketonuric patients points out that virtually all non-phenylketonuric children of phenylketonuric mothers are MR. However, placing such women on a low phenylalanine diet seems an acceptable alternative to sterilization. PKU fathers apparently do not affect the mental development of their offspring. (No refs.) - *B. Berman.*

Bay Clinic
Coos Bay, Oregon

2733 **EBRAHIM, G. J.** Nutrition rehabilitation centers. A growing childcare activity in developing countries. *Clinical Pediatrics*, 10(6): 303-305, 1971.

Nutrition rehabilitation centers in developing countries are achieving significant results in treating children with clinical syndromes of protein-calorie malnutrition (PCM) such as kwashiorkor and marasmus, entities that interfere with physical and mental growth and cause a large percentage of pediatric admissions and deaths in those countries. These centers seek to cure PCM, instruct the mother in caring for and feeding the child after hospital discharge (the mother stays with the child during hospitalization), and influence the community or village where the mother lives. Rapport with the instructor is most significant in instructing the mother; 80 percent of mothers learn sufficiently well to merit discharge in a week. New knowledge is quickly forgotten, so follow-up visits are necessary. Rehabilitation of parents and child is essential for a permanent cure. (3 refs.) - *B. Berman.*

Hospital for Sick Children
London, England

2734 **CLOW, CAROL L.; READE, TERRY M.; & SCRIVER, CHARLES R.** Management of hereditary metabolic disease. *New England Journal of Medicine*, 284(23): 1292-1298, 1971.

Immediate harmful consequences of mutations responsible for various hereditary metabolic diseases can be obviated by using allied health personnel in patients' homes to provide careful management, biochemical monitoring, and evaluation of patient reaction to therapy. An in-home program in a dense urban area for 64 Ss with 10 varieties of genetic disease, with 2 allied-health technicians furnishing 95 percent of patient contacts and continuous counseling, has offset impaired cellular and metabolic activity, and significantly reduced hospitalization. Simple chemical tests in the home sufficed to monitor biochemical status. Mass screening has speeded up detection of these afflicted individuals and demonstrated the large numbers with hereditary metabolic ailments that require diagnosis, continuous counseling, and long-term therapy. The success of in-home treatment depends largely on the awareness and support of parents or guardians. (18 refs.) - *B. Berman.*

Montreal Children's Hospital
Montreal, Canada

2735 **HEFFERNAN, ANDREW G. A.** Glucose tolerance and insulin response in phenylketonuria. *New England Journal of Medicine*, 285(1): 57, 1971. (Letter)

In the study of glucose tolerance and insulin response in phenylketonuria (PKU), Ss were 6 females with PKU who were never treated with a low phenylalanine diet (serum phenylalanine level 24.6±3.32mg per 100 ml), and 6 females matched for age and mental deficiency who did not have PKU (mean blood phenylalanine less than 5 mg per 10 ml). Following similar unrestricted diets (containing 300 g of carbohydrate unchanged for at least 3 days) for all Ss, a 50 g oral glucose tolerance test was performed after an overnight fast of 12-14 hours. None of the Ss had an abnormal glucose tolerance test, and there was no statistically significant difference in mean glucose and insulin values of the 6 PKU and 6 control Ss. Mean glucose area and insulin area demonstrated a lack of statistically significant difference in the total amount of insulin secreted by either group whose glucose area is similar. Results indicated that constantly elevated levels of phenylalanine do not induce hyperinsulinism or cause islet-cell fatigue, and that there is no difference in glucose tolerance or insulin response between normal and PKU persons. (6 refs.) - *K. L. Woodworth.*

University College
Dublin, Ireland

2736 **APPEL, MICHAEL R.; & MARKOWITZ, ALFRED M.** Massive splenomegaly in Gaucher's disease. *Journal of the American Medical Association*, 217(3): 343-344, 1971. (Letter)

The largest spleen ever to have been removed surgically was removed from a 22-year-old woman with Gaucher's disease. The patient presented with a markedly protuberant abdomen, ecchymoses, hemoglobin level of 6.8 gm/100 ml, hematocrit value of 17.5%, and platelet count of 10,000/cu mm. Splenectomy was performed through a left thoracoabdominal incision and the immediate postoperative course was uneventful. Weight of the resected spleen was 8,910 gm, approximately 20% of the patient's preoperative weight of 106 lb. Four months postoperatively the hemoglobin level was 10.3 gm/100 ml and the platelet count was 162,000/cu mm. Approximately 1 1/2 years after splenectomy the patient died suddenly at home of apparent respiratory tract difficulty following a short illness. Autopsy revealed edematous lungs weighing 1,250 gm with Gaucher cell infiltrates in the alveolar walls and a liver weighing 4,500 gm which was approximately 70% replaced by Gaucher cells. (8 refs.) - *K. L. Woodworth.*

No address

2737 **LATHAM, MICHAEL C.; & COBOS, FRANCISCO.** The effects of malnutrition on intellectual development and learning. *American Journal of Public Health*, 61(7): 1307-1324, 1971.

Late manifestations of intellectual impairment seem causally related to malnutrition (especially protein-calorie) in early life. Studies in Africa, India, and South America have shown the asso-

ciation between malnutrition and adverse socio-economic conditions. Malnutrition has adverse effects on behavior, personality, and school performance. In Bogota, Colombia, a massive longitudinal study, utilizing new methodologies (siblings fitting certain criteria), 3 teams of experts (nutrition and health, social anthropology, psychology), and batteries of tests, is seeking to define the effects of malnutrition. A new hypothesis holds that poor performance of many previously malnourished children results not from pathological CNS alterations, but from a caloric deficiency that restricts a child's activities and learning opportunities. The Bogota study is comparing children's activity with a new method—SAMI (Socially Acceptable Monitoring Instrument)—of counting heart rate. Although there is no exact knowledge of the relationship between dietary lack and intellectual retardation, we should take action now to remedy nutritional deficiencies in children. (36 refs.) - *B. Berman.*

Cornell University
Ithaca, New York

2738 **GREEN, JOSEPH B.** Neurophysiological studies in Batten's disease. *Developmental Medicine and Child Neurology*, 13(4): 477-489, 1971.

Neurophysiological studies were performed on 20 cases of presumed or proven Batten's disease (juvenile amaurotic familial idiocy), which is characterized by progressive retinal disease, convulsions, and intellectual decline. A characteristic EEG pattern (spikes or spike and wave complexes, and high voltage 2-4 per second slow-wave bursts) was observed in most of the 19 Ss with abnormal EEGs. Decrease in amplitude of both paroxysms and background activity accompanied the progression of the disease in the majority of cases. Also observed were photic activation in 7 cases and an abnormal evoked cerebral potential to median nerve stimulation in 5 of 6 cases tested. The remarkably large potentials evoked on the EEG by peripheral stimulation (including photic activation) have not been reported previously in Batten's disease. Although high amplitude potentials, evoked by visual and peripheral nerve stimulation, are not found only in Batten's disease, these exaggerated responses may be of great value in diagnosis when supported by EEG and electroretinogram results. (39 refs.) - *K. L. Woodworth.*

Indiana University School of Medicine
Indianapolis, Indiana 46202

2739 **MENKES, JOHN H.; ANDREWS, JOHN M.; & CANCILLA, PASQUALE A.** The cerebroretinal degenerations. *Journal of Pediatrics*, 79(2): 183-196, 1971.

Reviewed are the cerebroretinal degenerations, a group of hereditary diseases involving progressive intellectual and visual deterioration, some of which are characterized by abnormal sphingolipid metabolism and deposition of lipid within cells of the central nervous system. Since the review is intended to assist in bedside diagnosis, the disorders are grouped according to clinical findings (rather than nature of stored material or underlying enzymatic defect). The table of lipid storage diseases outlined, in terms of storage material (brain), enzymatic defect, and clinical characteristics, 3 major groupings of disorders: those without marked visceromegaly, those with marked visceromegaly, and white matter degenerations. Infantile amaurotic idiocy (Tay-Sachs disease), most common of the gangliosidoses, is described in terms of clinical manifestations, pathology, biochemical pathology, and differential diagnosis. Also focused on are late infantile amaurotic idiocy (an autosomal recessive disorder like Tay-Sachs disease), the 5 entities which assume the clinical course of late infantile amaurotic idiocy (late infantile neurovisceral storage disease with curvilinear bodies, late infantile amaurotic idiocy with lipofuscin storage, generalized gangliosidosis, juvenile GM_2 gangliosidosis, Niemann-Pick disease Group C), and juvenile amaurotic idiocy (lipofuscin storage disease and juvenile dystonic lipidosis). Each of these clinically defined degenerative syndromes can be the result of a number of genetic disorders. (62 refs.) - *K. L. Woodworth.*

UCLA Center for the Health Sciences
Division of Pediatric Neurology
Los Angeles, California 90024

2740 **KRIEGER, INGEBORG; & MELLINGER, RAYMOND C.** Pituitary function in the deprivation syndrome. *Journal of Pediatrics*, 79(2): 216-225, 1971.

Responsiveness of the hypothalamic pituitary axis in infants and children with the deprivation syndrome (growth failure, developmental retardation, age-dependent behavior abnormalities) was investigated. Responses of serum growth hormone and plasma cortisol to insulin-induced hypoglycemia were measured (adrenocorticotropic hormone release measured by the metyrapone test and through determination of plasma cortisol rise 60 minutes after injection of insulin). Ss were 9 infants (6/12 to 2 8/12 years) with maternal deprivation syndrome and 7 children (3 5/12 to 10 2/12 years) with psychosocial (deprivation) dwarfism, all of whom had no organic disease causing their growth failure. Comparison was made with data from infants whose linear growth failure was due to undernutrition on an organic basis. Fasting serum growth hormone values were elevated in the infants and some children. Only in the children were pituitary growth hormone and adrenocorticotropic hormone release unresponsive to hypoglycemia (5 of 7 children), though some (3 to 5) had original normal or elevated serum growth hormone values and all had normal metyrapone tests. Controls had elevated fasting serum growth hormone. Insensitivity to hypoglycemia was unrelated to severity of linear growth failure and probably does not indicate growth hormone deficiency at the target organ level. Undernutrition appeared to be the cause of the elevated serum growth hormone in the experimental Ss. (30 refs.) - *K. L. Woodworth.*

Children's Hospital of Michigan
Detroit, Michigan 48202

2741 SPERLING, MARK A.; HENGSTENBERG, FAY; YUNIS, EDUARDO; KENNY, FREDERICK M.; & DRASH, ALLAN L. Abetalipoproteinemia: Metabolic, endocrine, and electron-microscopic investigations. *Pediatrics,* 48(1): 91-102, 1971.

Abetalipoproteinemia was detected in 2 male siblings aged 8 and 11 years who presented with short stature and severely retarded skeletal maturation. A systematic endocrine evaluation was carried out, since short stature has never been the presenting complaint in this disease and since the physical appearance of the siblings resembled hypopituitarism. Thyroid indices and growth hormone responses to arginine and insulin stimulation were normal. Urinary 17 hydroxycorticosteroid levels were in the low range of normal, cortisol production was 1 to 2 standard deviations below the mean, and urinary excretion of catecholamines and their major components was normal. Steatorrhea with abnormal laboratory results on plasma proteins, lipids, carotene, and vitamins A and E provided evidence of malabsorption. Due to the lack of significant endocrine findings and the presence of malabsorption, it was felt that chronic malnutrition had led to the short stature. A trial of a medium chain triglyceride-rich diet was suggested as possible therapy. (36 refs.) - *B. J. Grylack.*

Harbor General Hospital
Torrance, California 90509

2742 Further recommendations for PKU management—Part I. *Clinical Pediatrics,* 10(8): 16B-16C, 1971.

All newborn infants should be screened early for phenylketonuria (PKU), in order to find affected children at a time when intervention may prevent the ill effects of the disease, especially MR. A blood screening program for newborns complemented by later urine screening at appropriate times with appropriate age groups would appear to be the ideal program. In fullterm infants, the earliest time when blood phenylalanine levels may be elevated will be 24, 48, or 72 hours after the first ingestion of milk by the infant, depending upon circumstances. In premature infants, a test sample of blood should be taken at 1 to 2 weeks or at discharge from the hospital, whichever is earlier. With newborns who are fullterm siblings of PKU patients, cord blood should be tested and the blood tests repeated at 3 and 5 days of age, and the infants should be placed and kept on a low phenylalanine diet until the diagnosis is made or excluded. A fullterm infant with confirmed serum phenylalanine levels of 20 mg/100 ml or more but with normal blood tyrosine concentrations may be placed provisionally on dietary treatment, but the diagnosis should be clarified soon through analysis of urine for phenylpyruvic acid and for orthohydroxyphenylacetic acid and by periodic dietary rechallenging. (No refs.) - *B. J. Grylack.*

2743 BEUTLER, E.; PANIKER, N. V.; & TRINIDAD, F. The assay of red cell

galactokinase. *Biochemical Medicine*, 5(4): 325-332, 1971.

A procedure has been devised which permits relatively rapid assay of the red blood cell enzyme galactokinase in large numbers of blood samples. ^{14}C-1-galactose obtained from several different commercial sources was mixed with magnesium, hexokinase, and adenosine triphosphate (ATP) in an incubative mixture, was passed through a microgranular slurry column, and was then frozen. Red blood cell hemolysates were then mixed with the reaction mixture. The percentage of galactose phosphorylated increased in an approximately linear fashion. The reactivity of the unlabeled galactose solutions changed with time, because of mutarotation and formation of a keto sugar. Blood samples from 42 children aged 2 to 16 years were found not to have significantly higher enzyme activities than adults. There was no correlation between the activity of galactokinase in the red cells and galactose intake in 29 adults who were studied. The age of the red cell population did not appear to exert a major influence on red cell galactokinase activity. The assay of red cell galactokinase appears to be clinically valuable in patients with cataracts, due to the association with galactokinase defects. (12 refs.) - *B. J. Grylack.*

City of Hope Medical Center
Duarte, California 91010

2744 **BLAU, KARL; & EDWARDS, DAVID J.** A modified automated fluorometric method for the determination of tyrosine in blood and tissues, and its specificity. *Biochemical Medicine*, 5(4): 333-341, 1971.

In order to analyze tyrosine in blood and other tissues, a modified method of the automated fluorometric determination of tyrosine by the nitrosonaphthol reaction was devised as a part of experiments dealing with simulation of phenylketonuria in experimental animals. A linear relationship was found between tyrosine concentration and fluorescence over the ranges covered by the 2 highest settings of the sensitivity of the fluorometer. The procedure covers the range of tyrosine concentrations normally found in blood spot eluates from newborn infants. Raised tyrosine levels, as may occur from immaturity of liver enzyme systems, especially in premature infants, and the high tyrosine blood concentrations that are a feature of hereditary tyrosinemia would need to be determined at a lower sensitivity setting, or at the regular setting after dilution of the blood spot eluate. This method seems to be simpler, more rapid, and more sensitive than previous methods designed to seek the same results. (15 refs.) - *B. J. Grylack.*

University of North Carolina School of Medicine
Chapel Hill, North Carolina 27514

2745 **CAMPBELL, A. G. M.; ROSENBERG, LEON E.; SNODGRASS, PHILIP J.; & NUZUM, CLAUDE T.** Lethal neonatal hyperammonemia due to complete ornithine-transcarbamylase deficiency. *Lancet*, 2(7717): 217-218, 1971. (Letter)

Assays of urea-cycle enzymes in the liver of a male infant who suddenly lapsed into coma on the third day of life and died 33 hours later with striking hyperammonemia revealed the first reported instance of complete absence of activity of the liver enzyme, ornithine transcarbamylase (O.T.C.). Activity of the other urea-cycle enzymes was within the range in appropriate control livers. Enzyme absence probably accounted for the high levels of hyperammonemia, which is now considered in the differential diagnosis of feeding problems and coma in newborns. O.T. C. is apparently coded for by an X-chromosome gene, mutations of which may lead to partial enzyme deficiency in heterozygous females and total absence in homozygous males. (3 refs.) - *B. Berman.*

Yale University School of Medicine
New Haven, Connecticut

2746 **HUNT, MELANIE M.; SUTHERLAND, BETTY S.; & BERRY, HELEN K.** Nutritional management in phenylketonuria. *American Journal of Diseases of Children*, 122(1): 1-6, 1971.

By creating a balance between the essential amino acids in a commercial low-phenylalanine protein substitute (Lofenalac) and supplementary phenylalanine from natural foods, a dietary treat-

ment for phenylketonuria (PKU) is provided which allows PKU children to develop normally. Serum phenylalanine (PA) levels of 5-10 mg/100 ml may be maintained. Dietary habits of the family may be followed closely, although amounts of some foods may need to be limited. Lofenalac is prescribed in amounts needed to supply the child's protein requirements at a given age. This intake is balanced with natural food intake so as to supply a total PA requirement of 70-90 mg/kg at 1 mo of age, decreasing to 35 mg/kg at 2 yrs and 25 mg/kg at 10 yrs. Suggested intake of Lofenalac by age is stated in level tablespoons of dry powder (1.4 gm protein and 7.5 mg of PA per tablespoon). When treatment is started, a natural source of PA (cow's milk is the most suitable source) is added to the formula to prevent a precipitate decline in the serum PA level. The amount of added milk required to maintain the serum PA level between 5 and 10 mg/100 ml helps to determine the initial amount of PA to be allowed from natural foods. The diet must be continually monitored and adjustments made as necessary. Insufficient Lofenalac and excess PA from natural foods are likely to cause elevated serum PA levels. The same may result when the PA from natural foods is so restricted that the body must break down its own tissues to provide the needed PA. Over a 3-year period all children treated from the age of 2 mos or less have normal growth rates, both in weight and height, and are of normal intelligence. (6 refs.) - *S. M. Rucker.*

Children's Hospital
Cincinnati, Ohio 45229

2747 **LOPEZ, RAFAEL; & COOPERMAN, JACK M.** Glucose-6-phosphate dehydrogenase deficiency and hyperbilirubinemia in the newborn. *American Journal of Diseases of Children,* 122(1): 66-70, 1971.

When jaundice and hyperbilirubinemia (HBA) appear in the neonatal period, and there is no blood group incompatibility between infant and mother, glucose-6-phosphate dehydrogenase (G-6-PD) deficiency should be suspected, particularly in ethnic groups in which a high incidence of the deficiency has been demonstrated. Earlier studies with American Negroes (incidence of G-6-PD deficiency, 8-10%) indicate that the deficiency alone does not produce HBA in the infants. Ingestion of certain drugs and infection are common causes of hemolytic anemia. Prematurity, acidosis, hypoxia, and factors normally associated with newborn in addition to inability of red blood cells (RBC) deficient in G-6-PD to metabolize hydrogen peroxide (H_2O_2) may also cause acute hemolytic anemia in these infants. However, when neonatal appearance of jaundice and anemia was noted in Negro infants free of blood group incompatibilities, a high proportion were found to have a G-6-PD deficiency. While the majority of the enzyme-deficient infants will be free of difficulties, the deficiency carries a potential risk for development of HBA. This is borne out by the 4 cases studied (one Puerto Rican girl and 3 American negro boys) with HBA and no blood group incompatibilities between mother and infant. Neither mothers nor infants had received drugs associated with hemolytic anemia. Tests of infants and families revealed that the infants had G-6-PD deficiencies of the RBCs. Evidence of congenital transmission is present. All newborns with HBA not associated with blood group incompatibility should be routinely screened for G-6-PD deficiency. (31 refs.) - *S. M. Rucker.*

New York Medical College
New York, New York 10029

2748 **QAZI, QUTUB H.; & THOMPSON, MARGARET W.** Dermatoglyphics in congenital virilizing adrenal hyperplasia. *American Journal of Diseases of Children,* 122(1): 28-30, 1971.

This study was undertaken to determine if there is a correlation between abnormalities of dermatoglyphics and incidence of congenital virilizing adrenal hyperplasia (CVAH). Dermal ridge formation on palms, fingers, soles, and toes begins in the third fetal month and is complete by about the eighteenth week. Abnormal genetic and environmental influences at this critical phase of fetal development can cause permanent alterations in the dermatoglyphics. In CVAH, due to C-21-hydroxylase deficiency, abnormal secretion of the adrenal cortex also begins in the third fetal month, and the fetus is exposed to massive amounts of androgenic substances. Dermal patterns of 19 girls and 15 boys with CVAH were studied. Dermatoglyphic abnormalities were noted only in the girls with CVAH. Since similar abnormalities were not noted in the boys, it seems unlikely that the female abnormalities

were caused by homozygosity of the gene for 21-hydroxylase deficiency. A more likely explanation for the larger dermatoglyphic pattern size observed in the girls with CVAH is the increased growth rate from the third month, when the fetus becomes exposed to large amounts of androgenic steroids. The disparity in growth rate between males and females is thought to be due to a greater end-organ responsiveness of the female tissues. Male fetuses normally are exposed to androgenic substances from their testes, and additional quantities do not induce more rapid growth. (13 refs.) - *S. M. Rucker.*

Downstate Medical Center
Brooklyn, New York 11203

2749 **HANAI, JUN; LEROY, JULES; & *O'BRIEN, JOHN.** Ultrastructure of cultured fibroblasts in I-cell disease. *American Journal of Diseases of Children,* 122(1): 34-38, 1971.

Ultrastructure studies of fibroblasts from 2 Ss with I-cell disease reveal that the inclusions in the fibroblasts are only weakly metachromatic compared with the intense metachromasia of the Hurler-Hunter inclusions and are filled with osmophilic membranous structures, whereas Hurler-Hunter inclusions are chiefly empty by electron microscopy. I-cell disease is characterized by abundant inclusion bodies within cultivated fibroblasts. It apparently is transmitted by an autosomal recessive trait. Features are extreme shortness of stature, maxillary hyperplasia, progressive mental and motor retardation, and bony deformities of the Hurler-Hunter type. Unlike the Hurler-Hunter syndrome, mucopolysaccharidurla is not present, nor is hepatosplenomegaly. Fibroblasts were taken not only from the 2 Ss, but from the parents of one and from 6 normal persons for control purposes. After being cultured and subcultured, the fibroblasts were appropriately prepared and examined by both light and electron microscopy. Occasional swollen hypergranular cells were found in the cultures from the father of the one patient, but not in those from the mother. Although occasional lysosome-like bodies or phagocytic vacuoles were seen in fibroblasts from controls, they did not have the complex morphology of the inclusions in I-cell fibroblasts. The location and morphology of the inclusions in I-cell disease suggest that it may be a lysosomal storage disorder. Further study is needed to determine if inclusions are present in cells *in vivo* as well as *in vitro.* (8 refs.) - *S. M. Rucker.*

*University of California
La Jolla, California 92037

2750 **PORTER, MYNA T.; FLUHARTY, ARVAN L.; & KIHARA, HAYATO.** Correction of abnormal cerebroside sulfate metabolism in cultured metachromatic leukodystrophy fibroblasts. *Science,* 172(3989): 1263-1264, 1971.

Metachromatic leukodystrophy (MLD) is a hereditary neurological disorder of lipid metabolism in which there is progressive degeneration of the nervous system. The biochemical lesion has been identified as a deficiency of the lysosomal enzyme arylsulfatase A. The manifestations of the genetic disorder in the tissue culture model can be corrected by an exogenous source of arylsulfatase A. When MLD cells are cultured in medium supplemented with a preparation of arylsulfatase A, the enzyme is incorporated by the cells, and the functional activity of the enzyme is retained over an extended period. When these cells are exposed to sulfatides, uptake and hydrolysis patterns characteristic of normal cells are observed. Sulfatides taken in by cultured MLD cells are found in structures that seem to be lysosomal, providing a basis for presuming that exogenously provided sulfatide and arylsulfatase A are taken into lysosomes by pinocytosis. The concept of enzyme replacement therapy is therefore feasible, and cultured fibroblasts serve as a useful model. (8 refs.) - *T. C. Turner.*

Pacific State Hospital
Pomona, California 91768

2751 **DAWSON, DAVID M.** Absence of guanine deaminase from cerebellum. *Neurology,* 21(6): 621-626, 1971.

A survey of the known enzymes of guanine metabolism in cat brain on a regional basis was undertaken as a result of the discovery that the Lesch-Nyhan syndrome is associated with absence of 1 of the enzymes in the salvage pathway of purine metabolism. Of the enzymes studied, guanine deaminase was the only enzyme with a

striking regional distribution. None could be demonstrated in the cerebellum. The metabolic role of guanine deaminase is obscure, since levels of free guanine in the brain, as elsewhere, are probably very low. It is possible that a regulatory function for this enzyme exists in the salvage pathway. It has been shown in boys affected with Lesch-Nyhan syndrome that the enzyme which acts on hypoxanthine and guanine is absent from tissues at autopsy. It is unclear how the findings concerning guanine deaminase distribution relate to that deficiency. (19 refs.) - *B. J. Grylack.*

Peter Bent Brigham Hospital
Boston, Massachusetts 02115

2752 **RIDDELL, A. G.** The hepatic glycogenoses. *Lancet*, 2(7715): 101, 1971. (Letter)

Operations of portacaval anastomosis or portacaval transposition can be performed in severe cases of type I or III glycogenosis and are recommended in children who show progressive deterioration on medical treatment alone. (1 ref.) - *B. J. Grylack.*

Bristol Royal Infirmary
Bristol, England

2753 **SPRANGER, JURGEN W.; GILBERT, ENID F.; TUFFLI, GORDON A.; ROSSITER, FRANCIS P.; & OPITZ, JOHN M.** Geleophysic dwarfism—a "focal" mucopolysaccharidosis? *Lancet*, 2(7715): 97-98, 1971. (Letter)

Three Ss presented with geleophysic dwarfism, which is characterized by small stature, a peculiar but pleasant and even happily natured facial appearance, a dysostosis-multiplex-like bone dysplasia affecting primarily the hands and feet, and an apparently "focal" accumulation of acid mucopolysaccharides (A.M.P.S.) in liver and possibly in cardiovascular tissue. Psychomotor development is slightly retarded at birth, and progressive growth failure and recurrent, severe respiratory infections are noted after the first year of life. The phenotype of these Ss does not fit any currently recognized mucopolysaccharidosis or mucolipidosis. The mesenchymal changes, in particular the skeletal involvement and joint contractures as well as the accumulation of A.M.P.S. in liver and cardiovascular tissues, justify the tentative categorization of this disorder as a mucopolysaccharidosis, although the apparently "focal" distribution of these changes differs from that seen in the "generalized" mucopolysaccharidoses. Geleophysic dwarfism, which affects both sexes, appears to be due to the homozygous state of a recessive gene, and the parents of these Ss have been given a provisional 25% recurrence risk. (5 refs.) - *B. J. Grylack.*

Universitats-Kinderklinik
Kiel, Germany

2754 **SUZUKI, YOSHIYUKI; JACOB, J. C.; SUZUKI, KINUKO; KUTTY, K. M.; & SUZUKI, KUNIHIKO.** G_{M2}-gangliosidosis with total hexosaminidase deficiency. *Neurology*, 21(4): 313-328, 1971.

The third known case of cerebral G_{M2}-gangliosidosis with total hexosaminidase deficiency has been reported, and clinical, morphological, and biochemical data have confirmed most of the findings in the original case and have provided additional information. The clinical course of this Caucasian, non-Jewish female was typical of classical Tay-Sachs disease, with progressive psychomotor deterioration, primary optic atrophy, cherry-red spots, marked startle response, and myoclonic and generalized seizures, with death at age 2 years 9 months. Histopathologically, the lesions in the brain could not be distinguished from those in classical Tay-Sachs disease. The important biochemical findings which distinguish this disease from Tay-Sachs are the additional accumulation of globoside in visceral organs and the total absence of both components A and B of hexosaminidase, as compared with an absence of component A alone in Tay-Sachs. (39 refs.) - *B. J. Grylack.*

University of Pennsylvania School of Medicine
Philadelphia, Pennsylvania 19104

2755 **WITZLEBEN, C. L.; SMITH, K.; NELSON, J. S.; MONTELEONE, P. L.; & LIVINGSTON, D.** Ultrastructural studies in late-onset amaurotic idiocy: Lymphocyte inclusions as a diagnostic marker. *Journal of Pediatrics*, 79(2): 285-293, 1971.

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Laboratory examinations of a 9-year-old Caucasian boy whose disease clinically and pathologically was a type of late-onset amaurotic idiocy included light and electron microscopic studies of peripheral white blood cells and a cerebral biopsy specimen (cerebral cortex and white matter). Found in the brain were a variety of lipid inclusions, including both "multilamellar cytosomes" and "fingerprint" inclusions, which suggests that these 2 morphologic types of inclusions may not always be indicative of separate metabolic disorders. Found in the lymphocytes were ultrastructurally striking lamellar cytoplasmic inclusions, which have not previously been described in this disorder (they appeared more distinctive than the granulocytic granules seen in this condition). Presence of these inclusions, identified by electron microscropy, may provide a distinctive and relatively easily observable diagnostic sign of the type of late-onset amaurotic idiocy present in this patient. (14 refs.) - *K. L. Woodworth.*

Cardinal Glennon Memorial Hospital
St. Louis, Missouri 63104

2756 **FIDLER, STUART M.; O'ROURKE, ROBERT A.; & BUCHSBAUM, H. WILLIAM.** Choreoathetosis as a manifestation of thyrotoxicosis. *Neurology*, 22(1): 55-57, 1971.

A thyrotoxic patient was observed with choreoathetoid movements which gradually receded and disappeared as the patient became euthyroid. The S, a 14-year-old Negro girl, was hospitalized with clinical signs of thyrotoxicity and choreoathetosis. The protein-bound iodine was 20 μg%, the T_3 uptake was 51%, and the thyroxine-T_4 by column was 19.6 μg% before propylthiouracil (PTU) therapy. After 15 days of therapy with 900 mg PTU daily, the thyroid studies of the patient were only mildly elevated, and her clinical signs of thyrotoxicosis and choreoathetosis were diminished considerably. After 75 days of therapy, she was clinically euthyroid and devoid of choreoathetotic movements, and her thyroid studies were normal. There was no evidence in this case of any of the more common causes of choreoathetosis. (13 refs.) - *B. J. Grylack.*

District of Columbia General Hospital
Washington, D. C.

2757 **WONG, PAUL W. K.; BERMAN, JULIAN L.; PARTINGTON, MICHAEL W.; VICKERY, S. K.; O'FLYNN, MARGARET E.; & HSIA, DAVID Y. Y.** Glutamine in PKU. *New England Journal of Medicine*, 285(10): 580, 1971. (Letter)

Plasma phenylalanine and glutamine concentrations were measured in 5 different groups of Ss: classic phenylketonuria (PKU) with retardation, untreated; classic PKU without retardation, untreated; classic PKU, treated; hyperphenylalanemic variant, untreated; and controls. Untreated classic PKU Ss had lower glutamine levels than normal controls. No significant difference in plasma glutamine was found between untreated Ss with classic PKU and normal intelligence and those with SMR. Instead, plasma glutamine was inversely related to plasma phenylalanine levels, with a correlation coefficienct of 0.57 ($t=3.6$, $p<0.01$). Results do not support the hypothesis, reported by another study, that lowered intelligence in PKU is due to a chronic deficiency of glutamine during the period of rapid brain growth. (4 refs.) - *K. L. Woodworth.*

Chicago Medical School
Chicago, Illinois

2758 **LEROY, JULES G.; SPRANGER, JURGEN W.; FEINGOLD, MURRAY; OPITZ, JOHN N.; & *CROCKER, ALLEN C.** I-cell disease: Clinical picture. *Journal of Pediatrics*, 79(3): 360-365, 1971.

I-cell disease, or mucolipidosis II, was the diagnosis for 8 patients having a hereditary syndrome characterized by early psychomotor retardation, severe shortness of stature, slowly progressive changes in facial and bodily configuration (a "Hurler"-like body configuration), and fatal outcome. Hypothesis of autosomal recessive mode of inheritance was supported by familial occurrence in 2 instances and parental consanguinity in another. Cultured fibroblasts showed an abundance of peculiar cytoplasmic granular inclusions (I cells), numerous deficiencies in the levels of the major lysosomal hydrolytic enzymes, and increase in lipid and mucopolysaccharide content. Urinary excretion of acid mucopolysaccharides was normal. Pathogenesis of the syndrome remains unknown, but the enzymatic and tissue chemical analytic data

suggest a storage disease, presumably centered in the combined lipid and mucopolysaccharide ("mucolipidosis") area. Of major concern in the identification of the I-cell disease patient is differentiation of the disease from Hurler's disease, G_{M1}-gangliosidosis type 1, and lipomucopolysaccharidosis. (18 refs.) - *K. L. Woodworth.*

*300 Longwood Avenue
Boston, Massachusetts 02115

2759 **TONDEUR, M.; VAMOS-HURWITZ, E.; MOCKEL-POHL, S.; DEREUME, J. P.; CREMER, N.; &*LOEB, H.** Clinical, biochemical, and ultrastructural studies in a case of chondrodystrophy presenting the I-cell phenotype in tissue culture. *Journal of Pediatrics*, 79(3): 366-378, 1971.

A Moroccan boy, hospitalized at 1 year of age, showed early and severe clinical and radiological signs of chondrodystrophy without increased mucopolysaccharidurla. Features permitting the case to be considered as a chondrodystrophy with disturbance in mucopolysaccharide metabolism include facial dysmorphy, joint stiffness, SMR, x-ray abnormalities, and vacuolized lymphocytes. There was also a significant decrease in liver β-galactosidase activity and an abnormal accumulation of $S^{35}O4$ in cultured fibroblasts. Ultrastructural studies showed clear membrane-bound inclusions particularly numerous in fibrocytes and glomerular epithelial cells. Only a few pleiomorphic inclusions were found in hepatocytes and neurons. Skin tissue culture showed cells filled with dark granules having an aspect characteristic of the I-cell phenotype. Many of the findings were quite peculiar, distinguishing this clinical pattern from the typical mucopolysaccharidoses. (41 refs.) - *K. L. Woodworth.*

*Hopital Universitaire St-Pierre
320, Rue Haute, 1000 Brussels, Belgium

2760 **PROCOPIS, PETER G.; &*TURNER, BRIAN.** Iminoaciduria: Benign renal tubular defect. *Journal of Pediatrics*, 79(3): 419-422, 1971.

At approximately 7 weeks of age, 200,000 healthy Australian infants were screened for abnormal aminoaciduria. In 170 cases, excessive excretion of proline, hydroxyproline, and glycine (iminoglycinuria) was found. Among the 170 infants with iminoaciduria, iminoglycinuria persisted in 15 cases and 22 cases developed a persistant hyperglycinuria. No clinical abnormality was found in the children in the series, except for 1 infant with mildly elevated blood pressure. The study confirms suggestions that iminoglycinuria of renal origin is a benign disorder or variation not associated with any recognized clinical abnormality (previous reports had associated iminoaciduria and a number of clinical manifestations, particularly MR). The heterozygote may be detected by an increased urinary glycine excretion, but not all heterozygotes manifest hyperglycinuria. (15 refs.) - *K. L. Woodworth.*

*Oliver Lathan Laboratory
New South Wales, 2113, Australia

2761 **DAWSON, GLYN; MATALON, REUBEN; & STEIN, ARTHUR O.** Lactosylceramidosis: Lactosylceramide galactosyl hydrolase deficiency and accumulation of lactosylceramide in cultured skin fibroblasts. *Journal of Pediatrics*, 79(3): 423-429, 1971.

Skin and bone marrow fibroblasts were cultured from a 3-year-old Negro female with lactosylceramidosis, a neurovisceral lipid storage disease characterized by accumulation of lactosylceramide and deficiency of lactosylceramide galactosyl hydrolase. Lipid analyses had revealed the specific accumulation of a lactosylceramide (galactosyl [1->4]glucosylceramide; GL-2) in a number of tissues and a marked galactosyl hydrolase deficiency in liver tissue; it was hypothesized that the defect would manifest itself either chemically or enzymically in tissue culture. The GL-2 defect was demonstrated both chemically (a twofold elevation) and enzymically in the fibroblasts. The GL-2 galactosyl hydrolase activity of the fibroblasts was reduced to less than 15% of normal, though total β-galactosidase activity was normal. Fibroblasts were cultured from skin and marrow biopsies of the patient's parents, and biochemical studies on them indicated that the disease was inherited in an autosomal recessive manner. (27 refs.) - *K. L. Woodworth.*

University of Chicago Box 228
Chicago, Illinois 60637

2762 **KARAYALCIN, GUNGOR; ROSNER, FRED; & SAWITSKY, ARTHUR.** Sea-blue histiocyte syndrome in an octagenarian. *Lancet,* 2(August 7): 318, 1971. (Letter)

A case of the sea-blue-histiocyte syndrome, the twenty-fifth reported, was seen in an 83-year-old white Anglo-Saxon woman. The syndrome is usually characterized by hepatosplenomegaly, mild thrombocytopenia with or without purpura, and presence in the bone marrow and spleen of the characteristic histiocyte, containing granules which Wright-Giemsa stain causes to stain bright blue or blue-green. Classic sea-blue histiocytes were found in 4 separate bone marrow aspirates over a period of 1 year, though a liver-biopsy specimen did not show any abnormal histiocytes. Findings in this patient differ from the classic description of the syndrome in 3 ways: she is much older (except for a 60-year-old woman, all reported cases are under 40), she does not have thrombocytopenia, and although she has an enlarged liver, no liver dysfunction can be demonstrated. It is likely that there are 2 forms of this syndrome, hereditary and acquired, and that this patient probably has the acquired type. (21 refs.) - *K. L. Woodworth.*

Long Island Jewish Medical Center
New Hyde Park, New York

2763 **WOODHOUSE, N. J. Y.; DOYLE, F. H.; & JOPLIN, G. F.** Vitamin-D deficiency and primary hyperparathyroidism. *Lancet,* 2(7719): 283-286, 1971.

Two female patients, one of them MR, with normocalcemic primary hyperparathyroidism and showing raised serum-alkaline-phosphatase levels and radiological evidence of bone disease were given small oral doses of vitamin D for 11 and 16 months, respectively. During treatment with vitamin D, the raised serum-alkaline-phosphatase levels returned to normal, remineralization of bone occurred and the bone lesions healed, intestinal absorption of calcium increased, and serum calcium and phosphate rose. Estimated dietary intake of vitamin D had been normal. Deficiency of this vitamin may occur as a consequence of an increased secretion of parathyroid hormone, rather than causing the hyperparathyroidism. This implies an increased requirement for vitamin D in this disorder. (21 refs.) - *K. L. Woodworth.*

Royal Postgraduate Medical School
London W. 12, England

2764 **ROSENBLOOM, ARLAN L.; SAVORY, JOHN; & LONDONO, JAVIER H.** The half-hour synthetic 1-24 corticotropin test of adrenocortical reserve in children. *Journal of Pediatrics,* 79(3): 489-493, 1971.

Adrenocortical reserve in children can be rapidly assessed by stimulating the adrenal cortex with intravenous administration of synthetic 1-24 corticotropin. Normal controls (N=9, 6-12 years) had a mean base-line value of plasma 17-hydroxycorticosteroid of 13.0 (range 5.7-21.7) prior to saline injection. Mean level was identical 30 minutes after saline infusion. Mean basal level of plasma 17-hydroxycorticosteroid on the day of corticotropin infusion did not differ significantly. All 9 responded to corticotropin with an increment varying from 6.1 to 19.7 μg per 100 ml in 30 minutes. In 17 patients requiring adrenocortical evaluation, the synthetic corticotropin was a reliable stimulator of a rise in plasma 17-hydroxycorticosteroid levels. Among 5 patients with meningitis, 2 with a ninefold elevation of resting plasma levels did not respond to the corticotropin; the 3 with a fivefold elevation did respond with a further elevation of 17-hydroxycorticosteroid. Rather than indicating adrenocortical insufficiency, these results suggest the upper limits of response during stress. The adult criteria for normal response in the 30-minute corticotropin test should be modified to allow a smaller increment for individuals with a basal level above 15 μg per 100 ml. (17 refs.) - *K. L. Woodworth.*

University of Florida College of Medicine
Gainesville, Florida 32601

2765 **WALKER, BRYAN A.; MURDOCH, J. LAMONT; McKUSICK, VICTOR A.; LANGER, LEONARD O.; & BEALS, RODNEY K.** Hypochondroplasia. *American Journal of Diseases of Children,* 122(2): 95-104, 1971.

In the course of a survey of achondroplasia in which 148 cases were diagnosed, an additional group of 13 individuals with hypochondroplasia was distinguished, with 2 instances of 2 affected generations in a single family. The clinical findings in hypochondroplasia are short stature with a relatively long trunk and disproportionately short limbs. Hypochondroplasia appears to be transmitted as an autosomal dominant trait. MR, which has been shown to be associated with hypochondroplasia, seems to be a much commoner association of this disorder than of achondroplasia, in which intelligence has been described as normal or slightly substandard. Roentgenographic changes in hypochondroplasia are similar to but milder than those in achondroplasia and do not show any skull involvement. The differential diagnosis of hypochondroplasia in infancy and early childhood includes other conditions of short limbed dwarfism manifest at birth. (27 refs.) - *B. J. Grylack.*

Johns Hopkins University
Baltimore, Maryland

2766 **COOK, J. G. H.; DON, N. A.; & MANN, TREVOR P.** Hereditary galactokinase deficiency. *Archives of Disease in Childhood*, 46(248): 465-469, 1971.

A 5-day-old male infant was discovered to have galactokinase deficiency when he presented with galactosuria as part of screening studies for hyperbilirubinemia. The parents were unrelated and had no gypsy blood. Hereditary galactokinase deficiency is a rare, recessive inborn error of galactose metabolism which apparently produces no dramatic disease manifestations in the early weeks and months of life. Progressive cataract formation is a major danger of the defect and is thought to result from the biosynthesis and accumulation of galactitol, an osmotically active sugar alcohol derived from galactose. Because of the need for early diagnosis and the existence of a therapeutic dietary plan, the differential diagnosis of obscure hepatic and splenic enlargement in the young infant should include this enzymopathy. (16 refs.) - *B. J. Grylack.*

Royal Alexandra Hospital for Sick Children
Brighton, Sussex, England

2767 **BOLANDE, ROBERT P.; & TIMMONS, G. DEAN.** Hepatosplenomegaly with opisthotonus in an infant. *Journal of Pediatrics*, 79(1): 142-150, 1971.

A clinical-pathological conference was convened in order to discuss a 4-month-old Negro girl with infantile Gaucher's disease. Important clinical features in this case were normal head size, hepatosplenomegaly beginning early in the disorder, strong but intermittent episodes of hyperextension of the neck, normal eye grounds, and significantly increased muscle tone. The patient died 51 days after the onset of the initial symptoms of fever and vomiting had appeared. At autopsy, the most striking gross features were the enlarged liver and spleen. Histological examination revealed typical Gaucher cells in liver, spleen, adrenal medullae, lymph nodes, intestinal lamina propria, thymus, bone marrow, and central nervous system. (9 refs.) - *B. J. Grylack.*

Case-Western Reserve School of Medicine
Akron, Ohio

2768 **COUTTS, N. A.; & FYFE. W. M.** Classical and mild phenylketonuria in a family. *Archives of Disease in Childhood*, 46(248): 550-552, 1971.

Of the 4 children who had classical phenylketonuria (PKU) in a 5-child family, 3 were untreated, the outcome being normal intelligence in the eldest and moderate MR in the other 2. The youngest child had been diagnosed in the neonatal period and was placed on a low phenylalanine diet. The untreated sibling of normal intelligence had an IQ of 104, while the other 2 untreated siblings had blood phenylalanine levels of 20 mg per 100 ml. Most patients with mild PKU have been diagnosed in the newborn period as a result of mass screening by the Guthrie test. While some of these patients have been treated from early infancy with a low phenylalanine diet, the relatively good prognosis for intellectual attainment in untreated mild PKU has raised serious doubts concerning the need for an expensive and unpalatable low phenylalanine diet. (6 refs.) - *B. J. Grylack.*

Hawkhead Hospital
Paisley, Renfrewshire, England

2769 **JESKE, J.; HUBNER, H.; BLASZYNSKA-NAWROCKA, DANUTA; & PERLICJUSZ, VIOLETTA.** A family with late

infantile amaurotic idiocy (Jansky-Bielschowsky's disease): Report of three cases. *Journal of Mental Deficiency Research*, 15(Part 2): 96-101, 1971.

Late infantile type of amaurotic family idiocy was reported in 3 of 4 siblings in a Polish family. The 2 elder children, twins, manifested the first symptoms of Jansky-Bielschowsky disease between 18 and 24 months of age. After a characteristic disease course, the boy died at 5.5 years of age and the girl, 6 months later. The youngest boy showed normal development until the second year. At present he is living and displays very advanced deterioration of the central nervous system function, blindness, and immobility. Despite the occurrence of the disease in 3 siblings, there is no basis for stating that the mode of inheritance is other than autosomal recessive. (16 refs.) - *B. J. Grylack.*

Institute of Pediatrics, Medical Academy
Lodz, Poland

2770 **BERENBERG, WILLIAM; & KANG, ELLEN S.** The congenital hyperammonemic syndrome. *Developmental Medicine and Child Neurology*, 13(3): 355-361, 1971.

A 9-month-old girl who presented with persistent vomiting, respiratory difficulty, and recurrent severe ketoacidosis was found to have a congenital hyperammonemic syndrome. Ammonia, the toxic end product of amino acid metabolism, must be metabolized to urea but either of 2 enzymes in the urea cycle (carbamyl phosphate synthetase or ornithine transcarbamylase) may be deficient, resulting in accumulations of ammonia. A casein loading test confirmed the diagnosis. The patient was found to do relatively well on severe protein restriction but generalized hypotonia and developmental delay were continually observed. (15 refs.) - *E. L. Rowan.*

Harvard Medical School, Department of Pediatrics
Boston, Massachusetts 02115

2771 **JANCAR J.** Prader-Willi syndrome. *Journal of Mental Deficiency Research*, 15(1): 20-29, 1971.

Two females and 3 males who exhibited Prader-Willi syndrome evidenced MR, hypotonia, obesity, hypogonadism, and growth retardation. Two of the male patients were brothers, and one was a twin with a physically and mentally normal twin sister. Although over 100 cases of this syndrome have now been reported, only 3 familial cases have been included. The nature of the lesion involved in Prader-Willi syndrome has not been identified, and no effective treatment has been specified. The syndrome has points of similarity with 9 other syndromes. (22 refs.) - *J. K. Wyatt.*

Stoke Park Hospital
Bristol, BS16 IQU, England

2772 **RUNDLE, A. T.** The thyroid gland as a major causative factor in growth abnormalities of mentally retarded children. *Journal of Mental Deficiency Research*, 15(1): 51-60, 1972.

Examination of serum thyroxine-iodine, creatinine phosphokinase, cholesterol, and alkaline phosphatase in relation to linear growth, weight, and skeletal development in 54 MR girls and 73 MR boys (CA 4 to 18 yrs) revealed 15 cases of hypothyroidism. In the hypothyroid group, serum cholesterol was significantly elevated, alkaline phosphatase was significantly reduced, and 13 Ss were at or below the twenty-fifth percentile of normal height for age. Comparison of these data with those on 330 unselected males revealed 22 cases of hypothyroidism. However, these cases did not evidence elevated serum cholesterol or reduced serum alkaline phosphatase. Neither the adult nor the child group exhibited a significant increase in creatinine phosphokinase levels in the serum. (31 refs.) - *J. K. Wyatt.*

St Lawrence's Hospital
Caterham, Surrey, England

2773 **MOORE, ROBERT Y.** "Metabolic" neuropathies and myopathies in infancy and childhood. *Developmental Medicine and Child Neurology*, 13(3): 394-395, 1971.

Metabolic neuropathies and myopathies in the pediatric age group differ in etiology and incidence from those in adults. Neuropathies are part of metachromatic leukodystrophy and Krabbe's

disease and are most commonly found in collagen diseases in children. The metabolic disorders which are associated with neuropathy in adult life may cause functional impairment of the nervous system without overt symptoms in children. (5 refs.) - *E. L. Rowan.*

University of Chicago
Department of Pediatrics
Chicago, Illinois

2774 **PERCY, ALAN K.; & KABACK, MICHAEL M.** Infantile and adult-onset metachromatic leukodystrophy: Biochemical comparisons and predictive diagnosis. *New England Journal of Medicine*, 285(14): 785-787, 1971.

The results of comparison of specific activities of arylsulfatase-A (ARA) and cerebroside sulfatase (CS) in leukocytes, urine and cultured skin fibroblasts in 7 children with infantile metachromatic leukodystrophy (MLD) and in similar materials from dizygotic twins with adult MLD (A-MLD) are reported. Urine ARA activity was analyzed by a modification of the Austin method and leukocytes and fibroblast activity assayed. The determination of CS activity with the use of biologically synthesized S-sulfatide as substrate was achieved by a modification of the technic of Mehl and Jatzkewitz. Urinary sulfatides were measured from 24-hour collections by means of the colorimetric method of Kean. Clinical diagnosis of MLD was confirmed in each S; urinary ARA activity was deficient in each case. ARA activity was compared to CS activity in A-MLD and I-MLD leukocytes and fibroblasts, with identical deficiencies of ARA and CS being found in both forms of the disease. The findings indicate that "partial enzyme deficiency" does not explain age-dependent variants of MLD, but the data do establish the feasibility of diagnosis before clinical symptoms appear, which is essential for accurate genetic counseling, carrier detection and intrauterine detection of MLD. (17 refs.) - *T. C. Turner.*

Johns Hopkins University School of Medicine
Baltimore, Maryland 21205

2775 **ABLETT, J. G.; & McCANCE, R. A.** Energy expenditure of children with kwashiorkor. *Lancet*, 2(7723): 517-519, 1971.

Thirteen infants with moderately severe kwashiorkor showed metabolic rates below normal; 4 normal controls showed normal rates. Metabolic rates were measured while Ss were asleep (an open-circuit technique with ventilated plastic hood was used). A few weeks of treatment with daily caloric intakes within a range of 130-150 C. per kg. brought the rate up to normal. In untreated kwashiorkor, basal metabolic rate is low, and apparently the energy changes associated with protein and calorie deficiencies in children resemble those reported for animals and human adults. (27 refs.) - *B. Berman.*

Medical Research Council
Kampala, Uganda, Africa

2776 **HUIGING, F.** Hepatic glycogenosis. *Lancet*, 2(7723): 550, 1971. (Letter)

A 10-year study of 98 Ss (most with hepatic glycogenosis) showed that (1) 56 (all males, since the defect is X-linked) had the kinase defect (opposed to a total of 28 with glucose-6-phosphatase deficiency) and (2) many persons with the kinase defect are not recognized because it is generally thought all glycogen diseases are associated with neurological symptoms. Its detection, however, is very important, since, under stress, there may be neurologic damage. (11 refs.) - *B. Berman.*

University of Miami School of Medicine
Miami, Florida 33152

2777 **ZACHMANN, M.; VOLLMIN, J. A.; NEW, M. I.; & CURTIUS, H. Ch.** Congenital adrenal hyperplasia due to deficiency of 11β-hydroxylation of 17α-hydroxylated steroids. *Journal of Clinical Endocrinology and Metabolism*, 33(3): 501-508, 1971.

A female child with congenital adrenal hyperplasia is the youngest child ever diagnosed with an 11β-hydroxylase deficiency. First diagnosed at age 1 week, the case permitted following the natural course of the disease without treatment during the first 14 months of life and showed an

excessively high secretion rate of compound S, without increase of compound DOC. The influence of 3β-hydroxysteroid dehydrogenase activity of infancy on the steroid pattern resulting from 11β-hydroxylase deficiency was suggested by very high urinary THS levels occurring only after urinary loss of 16 α-hydroxypregnenolone. Impairment of the 11 β-hydroxylation of 17 α hydroxylated compounds was reflected in a very high S secretion rate and low normal rate of F. This indirectly suggested also the presence in the human adrenal cortex of two 11 β-hydroxylating systems. (46 refs.) - *B. Berman.*

University of Zurich
Zurich, Switzerland

2778 **TATESON, R.; & BAIN, A. D.** G_{M2} gangliosidoses: Consideration of the genetic defects. *Lancet*, 2(7724): 612-613, 1971. (Letter)

A child in whom G_{M2} gangliosidosis-type 2 was diagnosed after demonstration of absence of hexosaminidase in cultured fibroblasts has revealed the basic defect in the condition and its connection with the lesion in other G_{M2} gangliosidoses. A change in characteristics as well as specific activity of hexosaminidase suggests the mutation in G_{M2} gangliosidosis is a structural rather than a regulator gene; in type 1, the mutation seems to be at the structural locus for hexosaminidase and substitutes an amino acid that prevents synthesis of the A form. (7 refs.) - *B. Berman.*

Royal Hospital for Sick Children
Edinburgh, Scotland

2779 **BROWN, ELEANOR S.; & WAISMAN, HARRY A.** Mental retardation in four offspring of a hyperphenylalaninemic mother. *Pediatrics*, 48(3): 401-410, 1971.

Four EMR offspring born to a hyperphenylalaninemic female with plasma phenylalanine levels averaging 16 mg/100 ml have been observed, and biochemical and clinical findings were recorded. These children did not appear to differ markedly from other EMR children in height or learning ability. Three of the offspring had growth retardation. The mother had a typical phenylketonuria (PKU) phenylalanine loading curve, the father's was normal, and the children's differed only slightly from the norm. Treatment of a PKU mother with a low phenylalanine diet during pregnancy is possible but difficult, and its success has not been proven satisfactorily. Prevention of such pregnancies is desirable. Routine screening of urines of pregnant women is being carried out in some clinics with Phenistix, so that the women with positive tests can be identified and the degree of hyperphenylalaninemia can be confirmed by analysis. (43 refs.) - *B. J. Grylack.*

University of Wisconsin Medical Center
Madison, Wisconsin 53706

2780 **SACHDEV, K. K.; TAORI, G. M.; & PEREIRA, S. M.** Neuromuscular status in protein-calorie malnutrition. Clinical, nerve conduction, and electromyographic studies. *Neurology*, 21(8): 801-805, 1971.

Thirty children with the clinical features of protein-calorie malnutrition were studied clinically and with nerve conduction and electromyograms in order to determine their neuromuscular status. All the Ss were edematous upon admission to the hospital, 22 had skin and hair changes associated with kwashiorkor, 26 had mental changes, 23 had muscle wasting, and 21 showed generalized weakness. The average serum total proteins was 3.91 g%, and the mean serum albumin was 1.53 g%. Nerve conduction velocity was delayed in 12 Ss. The needle electromyogram was abnormal in 14 of the 18 patients studied. Muscle biopsies done in 3 Ss showed neurogenic atrophy. Nerve conduction and needle electromyogram studies appear to be simple, reliable, and helpful in delineating the neuromuscular status of sick, apathetic, and malnourished children in whom an accurate clinical assessment of motor and sensory function is not possible. (15 refs.) - *B. J. Grylack.*

Christian Medical College Hospital
Vellore-4, Tamil Nadu, India

2781 **DANIEL, WILLIAM A., JR.; MOUNGER, JOANNE R.; & PERKINS, J. CAMILLE.** Obstetric and fetal complications in folate-deficient adolescent girls. *American Journal of Obstetrics and Gynecology*, 111(2): 233-238, 1971.

A prospective study of 163 pregnant girls of low-income status aged 12 to 15 years was carried out to determine the folate dietary intake, blood folate values, and the incidence of obstetric and fetal complications in these Ss. It was determined that 52% of the pregnant girls took in less than 10% of the recommended daily allowance (RDA) of folic acid, and 90.4% ingested less than 50% of the RDA. However, only 17% of the whole blood specimens showed less than 19 nanograms (ng) of folic acid, the lower limit of normal. Of the 126 pregnancies which were followed to completion, 3 had spontaneous abortions, 10 gave birth to premature infants, and 1 infant was stillborn as a complication of placenta previa. No patient was recorded as having third-trimester bleeding, abruptio placentae, or malformations of the newborn infants. The incidence of prematurity was almost identical in the normal and folate-deficient groups. Folate deficiency does not seem to be a significant factor related to abortion, third-trimester bleeding, abruptio placentae, or prematurity in young adolescent primiparas. (12 refs.) - *B. J. Grylack.*

University of Alabama Medical Center
Birmingham, Alabama

2782 **BEARD, ALICE; CORNBLATH, M.; GENTZ, J.; KELLUM, MIKE; PERSSON, BENGT; ZETTERSTROM, ROLF; & HAWORTH, J. C.** Neonatal hypoglycemia: A discussion. *Journal of Pediatrics,* 79(2): 314-324, 1971.

The discussion presents the authors' varying opinions on questions related to the pathogenesis, clinical manifestations, and management of neonatal hypoglycemia. Covered are pathophysiologic factors present in various types of hypoglycemia, levels of blood glucose regarded as diagnostic of hypoglycemia in full-term and low-birth-weight infants, studies on the incidence of symptomatic and nonsymptomatic hypoglycemia in infants of diabetic mothers and in low-birth-weight infants, and symptoms which can be considered pathognomonic of hypoglycemia. Also detailed are the authors' emergency and continuing treatment regimens for symptomatic hypoglycemia (frequency of obtaining serum glucose concentrations, fructose versus glucose for I.V. infusions, value of oral treatment) and for asymptomatic hypoglycemia (when oral feedings are started, milk versus glucose, role of I.V. glucose, frequency of blood sugar determination, differential treatment for infants of diabetic mothers and low-birth-weight infants). In addition, discussants explain how to manage infants at special risk from hypoglycemia who have normal blood glucose concentrations and no symptoms, when to use glucagon, epinephrine, or diazoxide, whether anticonvulsant drugs or long-term phenobarbital therapy (for infants with hypoglycemic seizures as neonates) should be used, and the prognosis of symptomatic (with and without seizures) and asymptomatic hypoglycemia in infants of diabetic mothers and low-birth-weight infants. (26 refs.) - *K. L. Woodworth.*

No address

2783 **GROVER, WARREN D.; & NAIMAN, J. LAWRENCE.** Progressive paresis of vertical gaze in lipid storage disease. *Neurology,* 21(9): 896-899, 1971.

Two brothers are described who had lipid storage disease and paresis of vertical gaze: one had gradually increasing ataxia and dysmetria between 5 and 8 years of age; convergence of the pupils was absent, eye movements were limited to 10° above the horizontal, there was a poorly defined macula with diminished foveal light reflex and pigmentary granularity; the other had symmetrically impaired vertical opticokinetic responses, with absence of convergence; an increased amount of coarse pigment with impaired foveal reflex of the macula; foamy histiocytes in bone marrow. The progressive limitation of upward and downward gaze, impaired convergence and intact lateral gaze suggest that involvement of the pretectal area is responsible for the oculomotor abnormality. The diagnosis of a lipid storage disease is suggested by the progressive course, organomegaly, retinal changes and foamy histiocytes found in the bone marrow. A central nervous system defect of metabolic origin should be considered in the evaluation of paresis of vertical gaze in childhood. (14 refs.) - *T. C. Turner.*

St. Christopher's Hospital for Children
Philadelphia, Pennsylvania 19133.

2784 **KAPPELMAN, MURRAY; THOMAS, GEORGE H.; & HOWELL, R. RODNEY.**

Histidinemia in a Negro child. *American Journal of Diseases of Children*, 122(3): 212-214, 1971.

The first documented case of histidinemia in a Negro child is presented and the normal plasma amino acid values found among a control group are discussed. S was a 7-year-old Negro girl who was hyperactive and had articulation errors and speech distortion. At 1 year she had a convulsion with viral pharyngitis and otitis media. Her older sister showed hyperactive school behavior and was also studied. An increase in blood and urine histidine concentrations, skin histidase activity and sweat urocanic acid levels warranted a diagnosis of histidinemia. Treatment of the S consisted of family counseling, speech therapy and tutoring. A control group of normal Negro children was studied to see if there were any racial differences in plasma amino acid concentrations. The study failed to show a significant differential in levels of blood histidine when related to the child's age except for a minimal rise during the early adolescent years. Histidinemia is due to a reduced activity of the enzyme histidine deaminase and appears to be inherited as an autosomal recessive. Histidine is an essential amino acid for the infant and child, but does not seem to be required by the adult. The primary problem of the disease seems to be a speech disorder. States of histidinemia do exist which are not openly expressed. (9 refs.) - *T. C. Turner.*

University of Maryland School of Medicine
Baltimore, Maryland 21201

2785 **HACKETT, THEODORE N., JR.; HACKETT, ROBERTA G.; *BRAY, PATRICK F.; & MADSEN, JACK A.** Chemical detection of metachromatic leukodystrophy in disease and carrier states. *American Journal of Diseases of Children*, 122(3): 223-225, 1971.

The results of 2 family studies involving late infantile and juvenile metachromatic leukodystrophy (MLD) are presented. Two siblings with MLD were seen at different stages of their disease; clinical findings were similar and diagnostically typical (abnormal cerebrospinal fluid (CSF) level, toluidine blue stain of urinary sediment revealing chromatic granules). A 12 year old girl of the family exhibited seizures, and after death, metachromatic deposits were found in cortical neurons and white matter. An older sister began manifesting slow dementia at 11; now 31, she has mild spasticity in her lower extremities and sural nerve biopsy specimen shows metachromatic deposits. Patients with MLD have very small amounts of arylsulfatase A in their circulating leukocytes. The white blood cells of the carrier contain an amount of the enzyme which is between the patients' and controls' (normal) levels, suggesting demonstration of a gene-dose effect. (6 refs.) - *T. C. Turner.*

*University of Utah College of Medicine
Salt Lake City, Utah 84112

2786 **MARTIN, EDWARD S.; & *GRIFFITH, JOHN F.** Myoclonic encephalopathy and neuroblastoma. *American Journal of Diseases of Children*, 122(3): 257-258, 1971.

The case is presented of an 8-month old female with acute myoclonic encephalopathy associated with neuroblastoma. She had constant generalized myoclonus with continuous opsoclonic eye movements and trunkal ataxia with no head control. Results of studies (EEG, x-ray, brain scan, pneumoencephalogram) made during admission were all normal. The S was given various anticonvulsants without effect and corticotropin without improvement. One month later the VMA spot test was positive and a chest roentgenogram showed a neuroblastoma in the left posterior mediastinal area, which was resected surgically. Following resection, there was marked improvement in the myoclonus which subsided by the twenty-eighth day after surgery. During an 8-month follow-up, VMA test and roentgenograms have remained normal. The child is now a normal 20-month old infant. Very small tumors, undetectable by any technique, can induce fulminant central nervous system derangement. There may be other examples of myoclonic encephalopathy of this type erroneously excluded from further consideration because an obvious tumor was not initially identified. Flaccid trunkal and limb tone has been a common finding in those patients who subsequently were delayed in psychomotor development. (9 refs.) - *T. C. Turner.*

*Duke University School of Medicine
Box 29751
Durham, North Carolina 27706

2787 **EPSTEIN, CHARLES J.; BRADY, ROSCOE O.; SCHNEIDER, EDWARD L.; BRADLEY, ROY M.; & SHAPIRO, DAVID.** In utero diagnosis of Niemann-Pick disease. *American Journal of Human Genetics*, 23(5): 533-535, 1971.

Assay of sphingomyelinase activity in cultured amniotic fluid cells now permits prenatal diagnosis of Niemann-Pick disease and offers the chance of unaffected offspring for parents at risk. Diagnostic amniocentesis for demonstration of the enzyme is thus an important aspect of genetic counseling. *In utero* diagnosis of Niemann-Pick disease was made in a gravida presenting at the clinic who had previously borne a child with this disease. A therapeutic abortion was performed by hysterotomy. Absence of sphingomyelinase activity in the fetal brain and liver confirmed the diagnosis. (11 refs.) - *B. Berman.*

University of California
San Francisco, California

2788 **EHRLICH, R. M.** Hypoglycaemia in infancy and childhood. *Archives of Disease in Childhood*, 46(249): 716-719, 1971.

Hypoglycemia is only a symptom of some underlying process. Convulsions are the most common presenting symptom of the disorder at any point during childhood but particularly under 1 year of age. In these children, hypoglycemia, associated with hepatomegaly, is usually due to diseases of the liver; gastroenteritis, central nervous system disease and islet cell tumor of the pancreas are other possible etiologies in that age group. After 1 year of age, ketotic hypoglycemia, hypopituitarism, and adrenal insufficiency are additional causes of the disorder. The child with symptoms suggestive of hypoglycemia but in whom blood sugar values are normal, the child with symptoms of hypoglycemia in whom a low blood sugar was found once, and the child with no symptoms in whom a low blood sugar was found once represent 3 groups that pose special problems for the pediatrician. Glucose tolerance tests, insulin, growth hormone, cortisol, and serum electrolyte levels together with serial blood sugar determination are all utilized in diagnosis. Management is geared towards elimination of the underlying cause, if possible, and towards control of acute symptoms of hypoglycemia in order to prevent resulting secondary damage. (12 refs.) - *B. J. Grylack.*

Hospital for Sick Children
Toronto 101, Ontario, Canada

2789 **COLOMBO, J. P.** Plasma glutamine in a phenylketonuric family with normal and mentally defective members. *Archives of Disease in Childhood*, 46(249): 720-721, 1971.

Plasma glutamine analyses were performed in 2 SMR siblings with phenylketonuria (PKU), their mother, and her brother in an attempt to evaluate a possible link between glutamine deficiency and mental defects in untreated PKU. The mother and her brother both had PKU but had normal and borderline IQs, respectively. The concentration of glutamine in the 2 affected children was found to be within the 2 standard deviation (SD) limit of normal values. The lowest concentration was detected in the mother, who had the highest IQ in the family. In contrast to a previous report of glutamine deficiency in a MR untreated PKU patient, the present study indicated no relationship between glutamine deficiency and MR. (7 refs.) - *B. J. Grylack.*

University of Berne
Berne, Switzerland

2790 **RAITI, SALVATORE; & NEWNS, GEORGE H.** Cretinism: Early diagnosis and its relation to mental prognosis. *Archives of Disease in Childhood*, 46(249): 692-694, 1971.

A total of 141 cases of hypothyroidism diagnosed within the first 2 years of life was reviewed in order to evaluate the features which permitted diagnosis within the first 3 months and to compare the mental level achieved in this group with that of patients diagnosed later. It was apparent that symptoms of hypothyroidism were more prominent than the classical signs. The most constant symptoms were constipation, feeding problems, and lethargy or inactivity. Fourteen out of 19 cases in which treatment was begun within the first 3 months of life had IQs above 90. In 4 out of 5 with IQs below 90, treatment was initiated during the third month.

Only 33% of the patients first treated from 4 to 6 months of age and 40% of those first treated after 6 months of age had IQs above 90. The early diagnoses of cretinism were suggested, therefore, by recognition of cardinal symptoms and were then confirmed by laboratory tests. L-thyroxine is the drug of choice for the treatment of cretinism. (12 refs.) - *B. J. Grylack.*

University of Maryland School of Medicine
Baltimore, Maryland 21201

2791 **SONKSEN, PATRICIA M.; COTTOM, D. G.; & HARDEN, ANN.** The evolution of the EEG in two patients with maple syrup urine disease (branched-chain ketonuria). *Developmental Medicine and Child Neurology*, 13(5): 606-612, 1971.

Clinical and EEG studies were done over a 1-year period in 2 cases of maple syrup urine disease (MSUD) following early diagnosis and early dietary therapy. Towards the end of the first week of life, both babies gradually developed increasing lethargy, with loss of primitive reflexes, progressive and serious cardiorespiratory problems, seizures, bizarre postures, and abnormal movements. The plasma levels of branched-chain aminoacids were grossly elevated (greater than 40 mg%). Dietary restriction was initiated at the end of the first 2 weeks. At 1 year of age, both infants showed a moderate degree of mental and motor retardation with slight hypotonia of the limbs. No gross neurological abnormalities were present. When the babies were critically ill neonatally, the EEG was grossly abnormal and showed bursts of irregular activity (2.5 to 6 cycles/second) mixed with discharges reaching 350 microvolts in amplitude alternating with periods of near equipotentiality. Through appropriate treatment, EEG features matured satisfactorily, and proper development of rhythmic activity was attained by 1 year of age. (17 refs.) - *B. J. Grylack.*

Hospital for Sick Children
London W. C. 1, England

2792 **MONTAGU, ASHLEY.** Sociogenetic brain damage. *Developmental Medicine and Child Neurology*, 13(5): 597-605, 1971.

Insufficient attention has been paid to the class of brain damage of sociogenetic origin which is produced by malnutrition and is functionally expressed chiefly by deficits in behavior, learning ability, and intelligence. All the evidence suggests that during the long period of brain growth, it is probably the first 6 months that are the most crucial; during this time, the brain is in its most rapid growth phase. It has been found that amount of deoxyribonucleic acid (DNA) is significantly reduced in the brains of malnourished children, a fact indicating the presence of a significantly smaller number of brain cells. Other deficiencies in the environment of the infant combine with malnutrition to hinder his developmental potential. In turn, the combination of adverse prenatal and postnatal environmental factors produces a continuum of psychosocial and psychophysical casualty. (51 refs.) - *B. J. Grylack.*

321 Cherry Hill Road
Princeton, New Jersey 08540

2793 **PERKINS, RICHARD P.** Hydrops fetalis and stillbirth in a male glucose-6-phosphate dehydrogenase-deficient fetus possibly due to maternal ingestion of sulfisoxazole. A case report. *American Journal of Obstetrics and Gynecology*, 111(3): 379-381, 1971.

A case was reported of a Negro male glucose-6-phosphate dehydrogenase (G-6-PD)-deficient fetus who presented with hydrops fetalis and stillbirth, possibly as a consequence of maternal ingestion of sulfisoxazole. The mother had taken sulfisoxazole for a urinary tract infection 2.5 weeks before the fetus of 36 weeks' gestation was delivered. No other causes for the hydrops and anemia were found. The severity of hemolytic response in G-6-PD-deficiency disease to stimuli such as sulfonamides is determined by the nature of the gene inherited as well as by sex. Despite the frequency with which the gene must certainly be associated with pregnancy and its allied stresses, no reports have been received previously of fetal death due to maternal ingestion of a drug known to incite hemolysis. Stillbirths in the susceptible population should be evaluated in order to exclude the possibility of maternal G-6-PD deficiency and fetal disease, since 3 to 4% or more of American Negro women can be expected statistically to carry an affected male fetus. (12 refs.) - *B. J. Grylack.*

622 West 168th Street
New York, New York 10032

2794 **MANN, LEON I.; SOLOMON, GAIL; CARMICHAEL, ANDREW; & DUCHIN, SYBIL.** The effect of metabolic acidosis on fetal brain function and metabolism. *American Journal of Obstetrics and Gynecology*, 111(3): 353-359, 1971.

Experiments were conducted on 10 mixed-breed ewes and their fetuses in order to evaluate the effect of metabolic acidosis on fetal brain function and metabolism. In 2 of 10 experiments conducted early in the study, rapid infusion of lactic acid at a rate of 11.5 ml/min for 3 minutes resulted in the sudden appearance of an isoelectric EEG, bradycardia, and a marked fall in cerebral perfusion pressure and cerebral blood flow. The severe metabolic acidosis ($pH<6.8$) could not be corrected with the infusion of tris-(hydroxymethyl) aminomethane (THAM). In the remaining 8 experiments, in which the rate of lactic acid infusion was lowering the pH more slowly but still significantly to 6.857 ± 0.084 ($p<0.005$), THAM corrected the acidosis. There were no significant changes in carotid artery oxygen tension, carotid blood flow, heart rate, or cerebral oxygen consumption during the slower infusion. The slowing of the EEG, decrease in amplitude, and appearance of sharp waves were not corrected significantly by THAM infusion. The similarity of the results of the present experiments to those noted during hypoxia in terms of cerebral function and metabolism might be explained by the marked increase in lactate ion that occurred in both experimental conditions. (13 refs.) - *B. J. Grylack.*

Cornell University Medical College
New York, New York

2795 **KRUMDIEK, CARLOS L.** The rural-to-urban malnutrition gradient. *Journal of the American Medical Association*, 215(10): 1652-1654, 1971.

Malnutrition in rural areas forces rural populations into cities where they combine to form urban slums. Rural populations have poorer diets than their urban counterparts, due to a seasonal variation in caloric and protein intake and the deleterious effects of utilizing most of the land for cash crops. These factors produce more and more severe malnutrition in rural inhabitants. In some countries where the rural population is well nourished, migrations to urban areas are not found. The question of the importance of rural malnutrition relative to other factors in stimulating migration to the cities might well be studied in these countries. (17 refs.) - *S. Glinsky.*

University of Alabama Medical School
Birmingham, Alabama

2796 **OZSOYLU, SINASI.** Diagnosis of Wilson's disease. *New England Journal of Medicine*, 284(20): 1159, 1971. (Letter)

The copper level of liver cannot be considered the absolute diagnostic criterion for Wilson's disease, as a previous letter contends. Other researchers have shown that the copper levels found in both primary biliary cirrhosis and long standing large bile duct obstruction are within those levels found in Wilson's disease. An editor's note refutes this contention on the ground that there is never any difficulty in distinguishing these conditions from Wilson's disease on clinical and histological grounds. (2 refs.) - *S. Glinsky.*

Hacettepe University School of Medicine
Ankara, Turkey

2797 **LUGO, GUSTAVO; O'NEIL, LYNN; & CASSADY, GEORGE.** Carcass water, fat, and chloride in the fetal growth-retarded rat. *American Journal of Obstetrics and Gynecology*, 110(3): 358-361, 1971.

No significant differences were found in body composition of growth-retarded and normally grown rat fetuses. *In utero* growth retardation was caused in 18 rats by unilateral uterine artery ligation. Fetuses in the untouched horn were the controls. Determinations of carcass water, fat, fat-free dry solid, and chloride were made in 54 growth-retarded and 93 normally developed control rat fetuses. The findings showed no significant differences in these parameters between the 2 groups, in contrast to a previous study which found increased body water and diminished body fat in the retarded group. The latter study measured viscera intact in carcass, while the present study totally eviscerated the fetuses. These results suggest the possible significance of compositional changes primarily in abdominal viscera during malnutrition. (15 refs.) - *S. Glinsky.*

University of Alabama Medical Center
Birmingham, Alabama

2798 **SANDSTEAD, HAROLD H.; CARTER, JAMES P.; HOUSE, FAYE R.; McCONNEL, FREEMAN; HORTON, KATHRYN B.; & ZWAAG, ROGER VANDER.** Nutritional deficiencies in disadvantaged preschool children. *American Journal of Diseases of Children,* 121(6): 455-463, 1971.

There is a lack of correlation between Stanford-Binet test scores and other biochemical indices of nutritional status in disadvantaged preschool children. The Ss were 100 preschool children from the economically depressed area of Nashville, whose nutritional status was evaluated and correlated with Stanford-Binet scores. The findings included mild growth retardation, nonspecific physical abnormalities, and, in 8.7%, goiter. Ninety-six percent of the children showed decreased body stores of vitamin A; there were also deficiencies in iron, folic acid, and thiamine. Hospital oriented biochemical screening tests gave normal results. There were significant correlations between initial IQ, final IQ, and IQ difference after language education, all of which related inversely to age. Biochemical indices and IQ showed no correlation. Anthropometric measurements showed correlation between height and serum vitamin A and iron, and an inverse correlation between head:chest ratio and erythrocyte transketolase activity. Home environmental factors contributed to malnutrition. (18 refs.) - *S. Glinsky.*

Human Nutrition Research Laboratory
Grand Forks, North Dakota

2799 **HSIA, Y. EDWARD; LILLJEQVIST, ANNE-CH; & ROSENBERG, LEON.** Vitamin B_{12}-dependent methylmalonic-aciduria: Amino acid toxicity, long chain ketonuria, and protective effect of vitamin B_{12}. *Pediatrics,* 46(4): 497-507, 1970.

The observed protein intolerance and ketoacidosis in patients with methylmalonicaciduria are direct results of the primary block in methylmalonate catabolism, which vitamin B_{12} partially overcomes. The S was a young boy with methylmalonicaciduria and a vitamin B_{12} dependency. A diet of 5 gm of protein per kg body weight resulted in transient ketoacidosis, long chain ketonuria, and increased methylmalonicaciduria. Injections of 1000 μg/day of vitamin B_{12} immediately lowered urine methylmalonic acid by 70% and partially corrected the defect in propionate found in the patient's leukocytes. Also, the clinical toxicity of a valine load was abolished. These results suggest that patients with this disorder should respond to dietary protein restriction and/or vitamin B_{12} supplementation. (26 refs.) - *S. Glinsky.*

Yale University School of Medicine
New Haven, Connecticut

MEDICAL ASPECTS – Etiologic Groupings

New growths

2800 **VOORHEES, MARY L.** Neuroblastoma with normal urinary catecholamine. *Journal of Pediatrics,* 78(4): 680-683, 1971.

Two cases of neuroblastoma are reported in which the infants had paresis from spinal cord compression by primary neuroblastoma originating in the cervical dorsal nerve roots and ganglia; both had normal urinary excretion of catecholamines and metabolites. These findings are in contrast to most neuroblastomas which originate in the adrenal medulla or in the sympathetic ganglia and are associated with abnormally high excretion of these materials. The patients, 16- and 20-month-old Caucasian females, had progressive paresis. The younger infant was hypotonic, maintained a frog position, and had no spontaneous motion or abdominal reflexes. A myelogram showed a complete block at the junction of C_2 and C_3 and a tumor involving the region of the dorsal roots was removed, but the patient died at 2½ years despite cobalt-60 and drug therapy. The other S had abnormalities limited to the neuromuscular system and could not sit, stand, or walk unsupported. A myelo-

gram showed a lesion at the level of C_7 to T_1 involving both dorsal roots and dorsal root ganglia. This was removed, and the S was given cobalt-60 therapy. The normal excretion of catecholamines and metabolites suggests that the tumor cells were unable to synthesize dopamine and norepinephrine. These findings have important implications in the diagnosis and follow-up treatment in neuroblastoma cases in that analyses of catecholamines will not aid in distinguishing certain types of tumors or in the evaluation of the response to therapy in such cases. (1 ref.) - *M. S. Fish.*

Upstate Medical Center
Syracuse, New York 13210

2801 **IMASHUKU, SHINSAKU; LABROSSE, ELWOOD H.; JOHNSON, EUGENE M., JR.; MORGENROTH, VICTOR H., III; & ZENKER, NICOLAS.** Tyrosine hydroxylase in neuroblastoma. *Biochemical Medicine*, 5(1): 22-29, 1971.

In neuroblastoma, catecholamines appear to exert a "normal" feedback inhibition on tyrosine hydroxylase. Assay of tyrosine hydroxylase activity in two neuroblastoma tissues (one from the left adrenal area of a 22-month-old Negro infant and the other from the paravertebral area in a 2½-year-old Negro girl) was carried out by liquid scintillation counting, utilizing L-tyrosine-3,5-^{3}H in the incubation mixture, and activity was compared to that of bovine adrenal tyrosine hydroxylase. Findings indicated that tetrahydropteridine ($DMPH_4$) was required as a cofactor, and activity was stimulated by Fe_{++}, observations which were comparable to those reported for bovine and pheochromocytoma enzymes. The neuroblastoma enzyme was inhibited to the same extent as the bovine enzyme by catechol compounds and other inhibitors at concentrations of 2.5×10^{-5} to 5×10^{-4} M. This indicated that the neuroblastoma enzyme is normally susceptible to feedback inhibition, in contrast to the pheochromocytoma enzyme which is less sensitive to catechol inhibition. Both neuroblastoma and pheochromocytoma are associated with increased synthesis of catecholamines. Because of the feedback inhibition on tyrosine hydroxylase exerted by these amines in neuroblastoma, this high rate of synthesis must operate by a mechanism other than that involved in pheochromocytoma. (21 refs.) - *M. S. Fish.*

University of Maryland School of Medicine
Baltimore, Maryland

2802 Neuroblastomas: Some children are best left on their own. *Medical World News*, 12(18): 23, 25, 1971.

Vigorous treatment may do more harm than good in children with neuroblastoma who have multiple nodules in their skin and livers, and perhaps even malignant cells in their bone marrow, but who present with no X-ray signs of skeletal involvement. A review of such neuroblastoma cases indicates that these youngsters respond well to little or no treatment and have as good a chance of survival as those whose tumors are confined to a single organ or structure. While clinicians dealing with these cases can remove as much of the primary tumor as possible, irradiation or chemotherapy should be applied with caution, if at all. Vigorous treatment may even interfere with the very defense mechanism that could save the child. Spontaneous regression does not ensure that the nervous system has not been harmed, however. In several cases of occult neuropathies associated with an encephalopathy, it is suspected that a previous tumor may have caused the encephalopathy before regressing. Most of these patients were MRs. (No refs.) - *B. J. Grylack.*

MEDICAL ASPECTS — Etiologic Groupings

Prenatal influence

2803 **JANERICH, DWIGHT T.** Relationship between the influenza pandemic and the epidemic of neurological malformations. *Lancet*, 1(7710): 1165, 1971.

A retrospective study of the epidemic of anencephalus and spina bifida in the U.S. which peaked in the years 1929-32 reveals that the parents of these children were earlier affected by the pandemic of influenza in 1918. Statistical profiles of those affected in New York State from September to November 1918 show that prepubertal and pubertal females were particularly vulnerable to the influenza; these same females would have

been at the peak of their reproductive careers at the time of the greatest incidence of central nervous system defects, from 1929-32. While the biological mechanism possibly linking these 2 epidemics remains obscure, additional epidemiological studies are suggested which can further examine the validity of the hypothesis. (2 refs.) - *N. Mize.*

Birth Defects Institute
Albany, New York 12208

2804 **CARTER, M. P.** Epidemics of anencephaly and spina bifida. *Lancet*, 1(7712): 1303-1304, 1971. (Letter)

The 1940-41 epidemic of hydrocephalus reported in an earlier paper has been improperly referred to by critics as a "pseudoepidemic." In fact, this epidemic was not at all an artefact of classification changes in the Registrar General's office as has been charged, since these changes were not made until 1963. Additionally, subsequent evidence has been developed by other researchers to support the original contention that the reported peak in infant hydrocephalus deaths was a genuine phenomenon, not a statistical quirk. (2 refs.) - *N. Mize.*

No address

2805 **PALANT, DANIEL I.; FEINGOLD, MURRAY; & BERKMAN, MILTON D.** Unusual facies, cleft palate, mental retardation, and limb abnormalities in siblings — a new syndrome. *Journal of Pediatrics*, 78(4): 686-689, 1971.

The presence of SMR and motor retardation associated with short stature, unusual facial characteristics, cleft palate, and limb abnormalities in 2 female siblings appears to constitute a new syndrome. The Ss, products of normal pregnancies and deliveries, and with normal birth weights, had similar facial appearances with almond-shaped eyes with mongoloid slant, small epicanthal folds, bulbous tip of the nose, nasolabial prominence, and prominent cupid bow of the upper lip. One male sibling had a slight similarity in appearance, but the Ss had no similarity to the parents or to other family members. The family had no history of MR or birth defects. Both Ss were inst., one at age 7 and the other at age 2. Chromosome and urinary amino acid studies of each child were normal. No other reported familial syndromes include all of the findings in these patients. The syndrome is likely transmitted by autosomal recessive inheritance. (10 refs.) - *M. S. Fish.*

Boston Floating Hospital for Infants
and Children
Boston, Massachusetts 02111

2806 **SCOTT, C. RONALD; BRYANT, JEAN I.; & GRAHAM, C. BENJAMIN.** A new craniodigital syndrome with mental retardation. *Journal of Pediatrics*, 78(4): 658-663, 1971.

A new syndrome affecting 3 male siblings with MR, unusual facial features, syndactyly, abnormal dermatoglyphics, and minor skeletal abnormalities is described. All Ss were born following normal pregnancies. The characteristics which the 3 Ss had in common were IQ between 35 and 50, growth retardation, and hirsutism. The unusual facial features included brachycephaly without craniosynostosis, small mandible, small pointed nose, prominent eyebrows, long dark eyelashes, and a "startled" expression. Soft tissue syndactyly occurred between the second, third, and fourth fingers and between the second and third toes. The dermatoglyphic pattern and other physical characteristics of the father were normal, but the mother had unusual dermatoglyphics and also had soft tissue syndactyly between the second and third toes, as did the maternal grandmother; however, she did not share other features in common with the 3 children. Laboratory studies of blood, urine, cerebrospinal fluid, and chromosomes revealed normal values. The syndrome is probably genetic in origin and may be X-linked. (10 refs.) - *M. S. Fish.*

University of Washington School of Medicine
Seattle, Washington 98105

2807 **HALL, BRYAN D.; & SMITH, DAVID W.** The Cornelia de Lange syndrome (continued). *Pediatrics*, 47(6): 1091-1092, 1971. (Letter)

Evidence presented for a secure diagnosis of Cornelia de Lange syndrome in a recently reported case study does not appear to be definitive. While many of the nonspecific findings in this syndrome were reported for the S, the diagnosis is questioned because the infant studied had a normal birth weight and a head circumference which was not microcephalic at 21 months. A review of the English literature appears to support this doubt, as does the absence in the reported case of very thin lips, a frequent finding in this disorder. The reported failure to thrive may have been partially due to the severe tetralogy of Fallot and may, in turn, have given rise to the secondary hirsutism. (4 refs.) - *M. S. Fish.*

University of Washington School of Medicine
Seattle, Washington 98105

2808 **KOLIN, IRVING S.; SCHERZER, ALFRED L.; NEW, BERTRAND; & GARFIELD, MARY.** Studies of the school-age child with meningomyelocele: Social and emotional adaptation. *Journal of Pediatrics*, 78(6): 1013-1019, 1971.

A study of school-age children with meningomyelocele has indicated the value of psychiatric screening and timely intervention in identifying and aiding patients in need of help and of the usefulness of initial and long-term family support, particularly in terms of communication between physician and family, in modifying the psychopatholgy and improving adaptation of these patients. A group of 13 Ss, ages 7 to 11 years and with IQs ranging from 46 to 117 (WISC), were evaluated psychiatrically, physically, neurologically, and psychologically. All were assessed in terms of mental status (presence of anxiety, depression, denial, identification, and projection), and the parent most involved with the child also received psychiatric evaluation and assessment in terms of the relationship to the child (overprotection, rejection, wish for an additional child, religious feelings, and denial). Good to fair adaptation occurred for 7 children and 5 parents; divorce or separation occurred in 6 of the families. Findings indicated that social and emotional factors were stronger determinants of adaptation than severity of physical impairment. (8 refs.) - *M. S. Fish.*

United States Naval Hospital
Orlando, Florida 32813

2809 **ROGERS, S. CLIFFORD; & MORRIS, MICHAEL.** Infant mortality from spina bifida, congenital hydrocephalus, monstrosity, and congenital diseases of the cardiovascular system in England and Wales. *Annals of Human Genetics*. 34(3): 295-305, 1971.

A study of infant mortality from spina bifida, congenital hydrocephalus, monstrosity, and congenital diseases of the cardiovascular system has shown large and highly significant secular variations in rates, lending support to an environmental etiology for these disorders. Infant mortality data for these anomalies were obtained from Annual Reviews of the Registrar General for England and Wales (1848-1967), were analyzed statistically for significance, and were compared with other series. Two periods of epidemic prevalence of dysraphism occurred in the British Isles and one in the United States, indicating the unlikelihood of a viral etiology. The suggestion is advanced that a genetic susceptibility of embryos to an antimetabolite in food under varying conditions of preparation, preservation, or storage may be involved. (23 refs.) - *M. S. Fish.*

4 Clifton Road
Rugby, England

2810 **LOCHRIDGE, STANLEY; PASS, ROBERT; & CASSADY, GEORGE.** Reticulocyte counts in intra-uterine growth retardation. *Pediatrics*, 47(5): 919-923, 1971.

Determination of reticulocyte levels (measured as % of circulating erythrocytes) of 145 neonates between the time of delivery and 24 hrs thereafter disclosed that in 65 normally grown, gestationally mature infants, reticulocyte levels were 4.9%, compared to 8.8% for 52 normally grown, gestationally immature infants. The mean reticulocyte count in 28 gestationally mature infants with intrauterine growth retardation (IGR) was 4.5%, almost identical to values obtained for their gestational peers. The growth retarded group had a significant excess of females and Caucasians. No significant differences in respiratory distress or perinatal asyphyxia were observed between the normally grown and the IGR groups, and venous hematocrits were not significantly different. (24 refs.) - *M. S. Fish.*

University of Alabama Medical Center
Birmingham, Alabama 35233

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2811 **SCHERZER, ALFRED L.; & GARDNER, G. GAIL.** Studies of the school age child with meningomyelocele: I. Physical and intellectual development. *Pediatrics*, 47(2): 424-430, 1971.

Appropriate intervention can aid many children with meningomyelocele to develop satisfactorily and to attain the capacity to master major cognitive and social tasks. The Ss were 14 survivors of meningomyelocele who, in the beginning school years, underwent physical and neurologic examinations which showed that the general physical condition of 13 who had re-evaluation of renal, orthopedic, and neurologic status was good. Eleven were fully ambulatory; and eye abnormalities, hydrocephalus, and the need for an ileal conduit occurred in 6, 9, and 8 cases, respectively. A system to rate the degree of impairment indicated that 8 had mild to moderate impairment while the remaining 6 had severe physical impairment. Psychological evaluation of the Ss showed that 3 were mildly retarded, 3 were moderately retarded, and 8 had low to high average intelligence. The median IQ was 88. Six patients attended regular school programs, and 8 attended special classes for the physically and/or mentally retarded. (15 refs.) - *M. S. Fish.*

New York Hospital
New York, New York 10021

2812 **RUSSELL, N. A.; *HOFFMAN, H. J.; & BAIN, H. W.** Intraspinal neurilemoma in association with the Rubinstein-Taybi syndrome. *Pediatrics*, 47(2): 444-447, 1971.

The first known association of neurilemoma with the Rubinstein-Taybi syndrome is reported. The S, a 10-year-old male, had a number of the characteristics of the syndrome: beaked nose, high arched palate, broad thumbs and first toes, and peculiar dermal patterns. Because of progressive weakness of the legs, restricted neck movements, and inability to stand without support, the S was readmitted 3 years later. X-ray studies of the spine disclosed an enlarged intervertebral passage on the left between C_3 and C_4, and a myelogram showed an intradural extramedullary mass at C_3. Cervical laminectomy resulted in the removal of a firm, dumbell-shaped, grape-sized tumor arising from the motor root of C_4. The tumor was found histologically to be a neurilemoma. Recuperation of the S was uneventful, and he regained the use of his limbs. The case, in addition to other reported patients with this syndrome who have pyramidal tract signs, indicates the possibility of discovery of still other patients with this syndrome who will present with CNS tumors. The syndrome possibly should be grouped with Von Recklinghausen's disease. (13 refs.) - *M. S. Fish.*

*The Hospital for Sick Children
Toronto, 101, Ontario, Canada

2813 **TAN, CHARLOTTE; ETCUBANAS, ERLINDA; LIEBERMAN, PHILIP; ISENBERG, HAZEL; KING, OSCAR; & MURPHY, M. LOIS.** Chediak-Higashi syndrome in a child with Hodgkin's disease. *American Journal of Diseases of Children*, 121(2): 135-139, 1971.

The first known case of Hodgkin's disease developing in a child with Chediak-Higashi syndrome is reported. This syndrome is a rare, autosomal recessively inherited disorder characterized by oculocutaneous pigmentary dilution, large anomalous granulations in the leukocytes, and increased susceptibility to infection. The S, an 8-year-old female when examined, had the stigmata of Chediak-Higashi syndrome: eye involvements (strabismus, poor vision, nystagmus, photophobia, and ocular albinism), albinoitic fundi, and fair, silvery hair. She also had recurrent skin abscesses, otitis media and extrema, and anemia. Mild motor and MR and neuropathy were noted. The S was in the "dull normal" range of IQ (IQ 82). Hematologic studies showed large anomalous granules in the leukocytes from both peripheral blood and bone marrow aspirates. The cervical lymph nodes increased in size even after tonsillectomy and adenoidectomy which were performed after cervical adenopathy was noted and despite antibiotic treatment. Biopsy of a lymph node confirmed a diagnosis of Hodgkin's disease. The S was given intensive therapy (irradiation and chemotherapy) but developed lung parenchymal involvement. A male sibling of the S had a similar physical appearance, was SMR (IQ 17), and died in an inst for the MR at the age of 15. Postmortem examination disclosed hepatosplenomegaly, hemoperitoneum, pyelonephritis, and extensive amyloidosis. (16 refs.) - *M. S. Fish.*

Memorial Sloan-Kettering Cancer Center
New York, New York 10021

2814 **LAURENCE, K. M.; & TEW, B. J.** Natural history of spina bifida cystica and cranium bifidum cysticum; major central nervous system malformations in South Wales, Part IV. *Archives of Disease in Childhood*, 46(246): 127-138, 1971.

Followup of the 425 cases of spina bifida cystica and cranium bifidum cysticum born in a population of 850,000 (4.12 per 1,000) in South Wales between 1956 and 1962 should provide a baseline for comparison of future surgical results with this essentially untreated group. Twenty-five percent were stillborn, 13% died during the first week of life, 47% died later, and only 15% survived. Survivors with meningocele (N=18) show little physical disability, normal intelligence, and some emotional instability. The cases with myelocele (N=37) show marked physical impairment, a wide range of intellectual ability, and good emotional adjustment. Survivors with encephaloceles (N=26) show relatively little physical disability but are generally MR. In this series spina bifida was more frequent and more severe in females. (38 refs.) - *E. L. Rowan.*

Llandough Hospital
Penarth, Glamorgan CF6 1XX, Wales

2815 **BURLAND, W. L.; SIMPSON, K.; & LORD, JULIET.** Response of low birthweight infants to treatment with folic acid. *Archives of Disease in Childhood*, 46(246): 189-194, 1971.

A group of 10 infants of low birth weight (less than 1800 grams) was given a series of 14 injections of folic acid (total 1.4 mg), and folate deficiency was compared over 9 months with a control group which did not receive supplemental folic acid. The control group showed a significant deficiency in both serum and red cell folate levels, while the experimental group maintained values within the normal range. The control group did not show obvious megaloblastic anemia or low hemoglobin but did demonstrate an increase in hypersegmented neutrophils, which might suggest subclinical folate deficiency. Folic acid should be given prophylactically to low birth weight infants. (22 refs.) - *E. L. Rowan.*

Glaxo Laboratories
Greenford, Middlesex, England

2816 **CHAR, FLORENCE.** "Turner phenotypes" vs Noonan's syndrome. *Journal of the American Medical Association*, 216(4): 679-680, 1971.

Since Noonan's syndrome appears to be a distinct entity with a dominant mode of inheritance and no relationship to Turner's syndrome other than some similar clinical features, its characteristics and the terminology used to describe it in the literature need clarification. Over 30 families with multiple cases of Noonan's syndrome have been reported. These individuals had normal 46 XY or 46 XX karyotypes. Congenital defects were widely varied, including congenital heart disease and distinctive facies of broad forehead and hypertelorism, flat nasal bridge, and thick, low-set ears. Among the features which may be variably present are webbing of the neck, short stature, cubitus valgus, proptosis, ptosis, wide-set nipples, proximal pectus carinatum, MR, cryptorchidism, abnormal dermatoglyphics, and abnormal electrocardiogram. Although frequently overlooked, Noonan's syndrome is often minimally expressed in the near relatives of affected individuals. (8 refs.) - *J. K. Wyatt.*

2817 **MILHORAT, THOMAS H.; & HAMMOCK, MARY K.** Isotope ventriculography. Interpretation of ventricular size and configuration in hydrocephalus. *Archives of Neurology*, 25(1): 1-8, 1971.

Examination of air and I 131 serum albumin ventriculograms in patients with hydrocephalus indicated that the pathological features of a particular case can influence the distribution of intraventricular isotopes and that interpretation of the results is complex and requires extensive experience. Of 9 hydrocephalic patients (7 with a severe condition) in which air and I 131 serum albumin ventriculograms were compared, precise conclusions about isotope distribution were difficult to make in 7 severe cases; however, in the 2 mild ones the evidence for rapid and extensive isotope migration into the brain parenchyma was convincing. The comparisons provided evidence for the conclusions that: a prompt and general mixing of the isotope with the cerebrospinal fluid (CSF) follows intraventricular injection of the isotope; the isotope dynamically demonstrates pathways of CSF circulation; the 2-hour ventricular scan after isotope injection produces a

dense inner zone (the contour of the ventricular system as defined by air) and a less dense outer zone corresponding to the isotope in the brain surrounding the ventricles; the eventual isotope distribution is complex and depends on the pathological features of individual cases. (24 refs.) - *M. S. Fish.*

Children's Hospital
Washington, D.C. 20009

2818 **ARONSON, STANLEY M.; & ARONSON, BETTY E.** Clinical neuropathological conference. *Diseases of the Nervous System*, 32(5): 346-351, 1971.

Clinical analysis and autopsy of a 31-year-old Caucasian female revealed all the major signs needed for the diagnosis of familial dysautonomia (Reilly-Day syndrome). Major clinical features include decreased lacrimation, abnormal esophageal motility, somatic motor disturbances, and emotional lability. Especially notable are diminished taste resulting from the absence of tongue papillae, olfactory deficits, and decreased temperature sensibility, possibly indicating a more generalized peripheral sensory deficit. Most families with this syndrome emigrated during the 19th century from a region in eastern Europe, including northern Moldavia, western Ukraine, and the city of Lvov. On the basis of identified cases it is possible to estimate the number of carriers in the U.S.; New York City alone houses nearly 25,000 asymptomatic carriers. (2 refs.) - *B. Berman.*

The Miriam Hospital
Providence, Rhode Island 02906

2819 **BROCKLEHURST, GORDON.** The pathogenesis of spina bifida: A study of the relationship between observation, hypothesis and surgical incentive. *Developmental Medicine and Child Neurology*, 13(9): 147-163, 1971.

Historically the description of spina bifida and hypotheses of its etiology have influenced the method of treatment. The hypothesis of total malformation due to primary hydrodynamic disturbances was supplanted by one of primary neuraxis malformation with secondary consequences. Theories of arrest, overgrowth, and traction have now given way to a teratological view. Teratogenic action at the site of neural tube closure affects depth, length, and tissue differentiation. Hydrocephalus results when this malformation compromises the flow of cerebrospinal fluid. Surgery within the first 24 hours of life is now accepted as the best means of preserving life and neurological function. (91 refs.) - *E. L. Rowan.*

University of Kentucky
Albert B. Chandler Medical Center
Lexington, Kentucky 40506

2820 **ATKINSON, W. J.** Studies in the movement of isotopes $RI^{131}SA$ and Na^{24} from ventricular CSF fluid. *Developmental Medicine and Child Neurology*, 13(1): 51-62, 1971.

A study of bulk movement of cerebrospinal fluid (CSF) in animals and humans, utilizing the injection of radioactive isotopes, is presented. Of 18 human cases of tension hydrocephalus in which Na^{24} or radioiodenated human serum albumin ($RI^{131}SA$) was injected into the right lateral ventricle or through the right frontal burr hole in the skull, radio scintillation counting showed that the 12 Ss who had one-half clearance times of 30 hours or more were cases of infantile hydrocephalus. In all but 2 Ss the half-times for clearance were greater than those reported in another study. Of 24 rabbits, two groups of 6 received 2.5 μC of the Na^{24} and the RISA, respectively; the other 12 (control group) died when the solution was injected. In the live series, examination of autopsy tissues showed that sodium was taken up more readily in the ependymal walls and the choroid plexuses than into the cerebral cortex and the sagittal sinus. The opposite occurred for the RISA. Observations on 3 cases of severe tension hydrocephalus in children tended to bear out that even in these cases sodium passes through the ependymal walls readily, whereas RISA does not. In experiments in which internal hydrocephalus was caused in 6 otherwise normal rabbits, assay of tissues after injection of the radioactive isotopes showed little uptake of radioactive material. Shortest half-times for isotope clearance were recorded for a patient with bilateral subdural hematoma. After shunt installation in this S, clearance of both sodium and RISA from the head indicated satisfactory operation of the shunt. Comparison of

clearance times of the isotopes in Ss both before and after shunt operations indicated that clearance half-times were not reduced to very low figures. (5 refs.) - *M. S. Fish.*

Hurstwood Park Hospital
Sussex, England

2821 **TURNER, GILLIAN; TURNER, BRIAN; & COLLINS, EDITH.** X-linked mental retardation without physical abnormality: Renpenning's syndrome. *Developmental Medicine and Child Neurology*, 13(1): 71-78, 1971.

MR which is present in males without physical or other somatic abnormalities may be an X-linked disorder which should likely be designated as Renpenning's syndrome. The division of 148 moderately retarded males (IQ 36-51) into 2 groups was: Group I, 17 Ss in which 6 criteria were absent; and Group II, 131 Ss having one or more of these criteria. The criteria were: height and/or head circumference above the ninety-seventh percentile or below the third percentile; fits requiring continuous medication; one major or 3 minor congenital abnormalities; a definite diagnosis; and neurological signs. Examination of the Ss and their family histories indicated that among the 17 Group I Ss, 7 families had a similarly affected (retarded) brother or brothers who also fell into Group I, having none of the above criteria. Four other families had a history compatible with x-linkage. Of the 131 Group II Ss, 23 had an affected sibling (13 female and 20 male). The results show that of Group I males, 41% had a similarly affected brother but no affected sisters, whereas only 17% of the Group II males with abnormalities had affected siblings, and brothers were affected in only 8% of this group. Although a number of other X-linked diseases are associated with MR, biochemical or somatic stigmata are present in these, in contrast to Renpenning's syndrome, which likely accounts for approximately 10% of all moderately retarded males. (9 refs.) - *M. S. Fish.*

Grosvenor Hospital
Sydney, New South Wales 2130

2822 **VIDYASAGAR, D.; & CHERNICK, V.** Continuous positive transpulmonary pressure in hyaline membrane disease: A simple device. *Pediatrics*, 48(2): 296-299, 1971.

An improvement in oxygenation of infants with hyaline membrane disease (HMD) has been reported based upon a simple modification of the standard incubator-respirator which allows the application of a constant negative pressure (CNP) to the thorax of the infant. Five infants with HMD were studied using CNP by subjecting the body below the neck to a constant negative pressure of 5 cm H_2O. The infants ranged from 31 to 38 weeks in gestation and from 1.25 to 2.05 kg in birth weight. Following the application of CNP, there was a highly significant increase in arterial PO_2 which averaged 35 mm Hg (t_d=6.99, $p<.001$) and a decrease in alveolar-arterial PO_2 difference. There was no significant influence on arterial PCO_2. CNP appears to improve gas exchange in infants with HMD, since it prevents airway closure during expiration. The need for endotracheal intubation and artificial respiration may be decreased as a result of this treatment approach. (3 refs.) - *B. J. Grylack.*

University of Manitoba
Winnipeg 3, Manitoba, Canada

2823 **SAHAR, A.; HOCHWALD, G. M.; KAY, W. J.; & RANSOHOFF, J.** Spontaneous canine hydrocephalus: Cerebrospinal fluid dynamics. *Journal of Neurology, Neurosurgery and Psychiatry*, 34(3): 308-315, 1971.

Bulk formation and absorption of cerebrospinal fluid (CSF) were studied in spontaneous hydrocephalus in 14 dogs with the aid of the ventricular perfusion technique. Of the 14 dogs, 7 had severe aqueductal stenosis, 2 had no detectable aqueduct, and 5 had communicating hydrocephalus. Histologically, the severest changes were observed in the lining ependyma and periventricular white matter. The ventricular pressure measured at the time of penetration of the ventricles ranged between 5 cm H_2O and 15 cm H_2O. The rate of formation of CSF decreased with perfusion pressure by V_f 0.02595+0.00022 P ml/min (P pressure in cm H_2O). The absorption of spinal fluid increased linearly with pressure by V_a 0.0165+0.00050 P. The finding of low ventricular pressures may bear a relationship to certain variants of human hydrocephalus.

Intraventricular pressure which may be considered well within normal range is not a rare finding even in uncompensated human hydrocephalus. The observation of a relatively low pressure in any patient may indicate an approximation to a new equilibrium, due to an increase in absorption and a decrease in formation of CSF. (25 refs.) - *B. J. Grylack.*

Hebrew-Hadassah Medical School
Jerusalem, Israel

2824 **McKUSICK, VICTOR A.** The nosology of genetic disease. *Hospital Practice*, 6(7): 93-101, 1971.

Accurate nosology (classification of disease) is essential for prognosis, understanding, and therapy, but for genetic disease it is of key importance in prophylaxis. Genetic nosology has progressed from 400 identified entities in 1958 to more than 800 today. Two fundamental phenomena the genetic nosologist must cope with are pleiotropism (multiple effects of a single mutant gene) and heterogeneity ("one from many" as against "many from one"). These phenomena are reflected in 2 chief tendencies among nosologists: "lumping" and "splitting." Accurate classification depends on 3 groups of evidence: clinical, genetic, and biochemical. At times, a "clustering" of cases can lead to identification of a hitherto occult kinship (e.g., amyloid neuropathy). Other kinds of evidence include mode of inheritance, genetic linkage, and matings (of identical autosomal recessive disorders). The most definite means of establishing genetic heterogeneity is through biochemical studies. A recent development involves the "correction factor," a diffusible substance (probably a group of enzymes) found in cell cultures and in normal human urine. (No refs.) - *B. Berman.*

Johns Hopkins University
Baltimore, Maryland

2825 **ROBINSON, CARL D.; PERRY, LOWELL W.; BARLEE, AMNAT; & MELLA, GORDON W.** Smith-Lemli-Opitz syndrome with cardiovascular abnormality. *Pediatrics*, 47(5): 844-847, 1971.

The fortieth reported case of the Smith-Lemli-Opitz syndrome, with associated congenital heart disease, was hospitalized at age 2 days for a heart murmur noted at birth. Examination revealed a pansystolic murmur and generalized cardiomegaly; cardiac catheterization revealed an incomplete endocardial cushion defect. After discharge the child failed to thrive; she had a low thyroid level and retarded bone age. She was treated with thyroid extract but expired of bilateral bronchopneumonia on her fourth admission to the hospital. The syndrome, described in 1964, shows failure to thrive, MR, microcephaly, neuromuscular disorders, and typical facies. Chromosomal karyotype is normal. Hypospadias (with or without cryptorchidism) affects 95 percent of male Ss. Of the 40 cases, 20 percent had associated cardiovascular anomalies; congenital heart disease is considered a common feature. (17 refs.) - *B. Berman.*

Children's Hospital
Washington, D. C.

2826 **CHEN, ANDREW T.L.; FALEK, ARTHUR; & LESTER, WILLIAM.** Full-term low birth-weight and chromosomes. *Lancet*, 2(7722): 496-497, 1971. (Letter)

Full-term low-birth-weight neonates show a higher incidence of chromosomal abnormalities than do controls and have a poor prognosis for survival. Chromosomal analysis of all full-term low-birth-weight newborns delivered at Georgia Baptist Hospital, Atlanta, Ga., since September 1, 1969 (202 out of 7,533) has shown 5 with chromosomal anomalies; in a matched control group there was one (stillborn) with a chromosomal anomaly. (4 refs.) - *B. Berman.*

Emory University
Atlanta, Georgia

2827 **OUNSTED, M.; & TAYLOR, M. E.** The postnatal growth of children who were small-for-dates or large-for-dates at birth. *Developmental Medicine and Child Neurology*, 13(4): 421-434, 1971.

Weight, height, and head circumference measurements were made at followup examinations of 106 small-for-dates (SFD) children (2 or more standard deviations below mean birthweight for their length of gestation) and 100 large-for-dates (LFD) children (2 or more standard deviations

above mean). SFDs were aged up to 5 years and LFDs up to 3 1/4 years. Mothers were interviewed, and their pregnancies and backgrounds were studied. Although at the latest examination Ss' weight was still deviant in the original direction (other 2 measures were also, but to a lesser degree), there was a general tendency to revert towards the population mean. Certain prenatal factors were weakly associated with the speed of such a reversion. Among the SFD Ss who grew most slowly after birth, there was higher incidence of placental and fetal pathology and of maternal hypertension and underweight. SFDs who were the firstborn of a well-built mother had the best chances of reverting rapidly to the mean. Ss whose small fetal size was due to the prepotency of a physiological maternal constraining factor grew faster after birth but did not reach the mean by school age. Among the LFDs who continued to grow fast, upper social classes were underrepresented, but associations between divergent postnatal growth patterns and LFD pregnancies were not found. Since individuals vary so much in growth and development, only long-term followup, investigating many developmental variables, can assess the varying effects of deviant fetal growth rate on postnatal growth. (29 refs.) - *K. L. Woodworth.*

Radcliffe Infirmary Department
of Obstetrics and Gynaecology
Oxford OX2 6HE, England

2828 **TELFER, M. A.; SUGAR, M.; JAEGER, E. A.; & MULCAHY, J.** Dominant piebald trait (white forelock and leukoderma) with neurological impairment. *American Journal of Human Genetics,* 23(4): 383-389, 1971.

Findings in 8 individuals in 2 families affected with the autosomal dominant piebald trait, which includes a white forelock and leukoderma, were studied in order to document further the nature of the phenotype. Since depigmentation of the skin and hair was congenital rather than acquired, vitiligo was excluded. The white forelock, leukoderma, medial whitening of the eyebrows, and hearing loss were consistent with Waardenburg's syndrome, but the broadened nose root, lateral displacement of the lacrimal ducts, and hypoplasia of the iris were not present. Although the Ss differ in some respects from individuals described as having typical piebald trait, the autosomal dominant inheritance, congenital white forelock, and patches of leukoderma, as well as the absence of the other features mentioned, suggested that this was the correct diagnosis. Atypical features found in these Ss included dorsal depigmentation of the trunk, presence of a parietal tuft of white hair in 1 individual rather than the typical white frontal blaze, neurosensory hearing loss, cerebellar ataxia, impaired motor coordination, and, in 7 of the 8 Ss, MR. (14 refs.) - *B. J. Grylack.*

Elwyn Institute
Elwyn, Pennsylvania 19063

2829 **BAUER, CHARLES H.; & TINKLEPAUGH, WENDY.** Low birth weight babies in the hospital. *Clinical Pediatrics,* 10(8): 467-469, 1971.

Liberalizing changes initiated in the Newborn Nursery at the Roosevelt Hospital in New York City proved successful and motivated the decision for early discharge of low birth weight patients, regardless of body weight, if a suitable home and parental competence in baby care were ensured and if the infant showed a satisfactory clinical condition. During the period December 1968 to June 1970, 57 low birth weight infants (range 885 to 2,400 g) were discharged at an average weight of 2,058 g. Follow-up of 53 patients showed that 51 (89%) were thriving and that 2 (4%) were doing poorly. One of these subsequently succumbed to E. coli septicemia, which apparently began 5 days following discharge from the hospital. The other infant grew slowly at first but is currently doing well at home. Of the 57 infants, 7 (12.3%) had a gestational age of less than 32 weeks; 22 (38.6%), 32 to 35 weeks; 24 (42.1%), 36 to 40 weeks; 3 (5.3%), over 40 weeks; and 1 (1.7%), unknown. Other factors related to the prenatal course of the mother and to the blood picture, sex, birth, and feeding of the baby were also studied, but none proved of any significance. Similar results obtained from follow-up for at least 2 months after discharge of infants under 5 lbs indicated that none had been ill. (8 refs.) - *B. J. Grylack.*

Roosevelt Hospital
New York, New York 10019

2830 Shunt tested in hydrocephalic youngsters. *Journal of the American Medical Association,* 217(2): 133-134, 1971.

A shunt which allows hydrocephalic children to swallow cerebrospinal fluid along with saliva through its termination in Wharton's duct of the submandibular gland has been tested in only 10 children, the longest follow-up being 1 year. One patient received a primary shunt, and 9 underwent standard shunting procedures plus at least 2 revisions each prior to insertion of the new shunt. Complications of bowing and skin erosion occurred in 2 cases, but otherwise the shunts operated successfully. There has been no evidence of retrograde infection from the mouth into the brain. The Wharton's duct shunt length will probably have to be altered less frequently than other shunts, because the rate of growth differential is less over the duct shunt's short course than over the distance of the atrial or peritoneal shunts. (No refs.) - *B. J. Grylack.*

2831 **CZEIZEL, ANDREW; & BOGNAR, ZOLTAN.** Mortality and morbidity of legal abortion. *Lancet*, 2(7717): 209-210, 1971. (Letter)

Analysis of 1,816,429 cases of legal abortion in Hungary showed a mortality rate of only 1.76 per 100,000 (32 maternal deaths). Legalization of abortion considerably decreased mortality and morbidity. However, induced abortion showed a high incidence of placenta praevia, premature placental detachment, Rh isoimmunization, and low-birth-weight babies. (1 ref.) - *B. Berman.*

National Institute of Public Health
and Maternal Health Insciitute
Budapest, Hungary

2832 **OUNSTED, MARGARET.** Biological factors and fetal growth. *Developmental Medicine and Child Neurology*, 13(4): 524-528, 1971.

Studies cited in the review of research on fetal growth rate demonstrate that the rate of growth of the human fetus varies greatly both between and within ethnic groups. Factors contributing to the wide variation in mean birthweight between ethnic groups may include parental stature, socioeconomic factors, parity, maternal nutrition and smoking, and geographic location. One study even related mean birthweights of North American Indians to their pre-Columbian living habits. Fetal growth rates can differ as widely within as between ethnic groups. Marked increases in the mean birthweight of infants in specific populations have been recorded. A maternal constraining mechanism which modulates fetal growth rate has been suggested. The set of the regulator may be determined by the degree of constraint imposed on the mother when she herself was a fetus. Transmission of constraint by this method would facilitate rapid changes in fetal growth rate, as different generations adapt to variations in living conditions. All major studies agree that the male conceptus grows faster than the female in singleton pregnancies. In conclusion, the studies indicate that there are, as yet, no real apprehension of the basic mechanisms regulating growth rate of the human fetus, and no sound explanations as to why there is so much variety between and within different ethnic groups. (35 refs.) - *K. L. Woodworth.*

Radcliffe Infirmary Department of Obstetrics
Oxford, England

2833 **PEARSON, J.; BUDZILOVICH, G.; & FINEGOLD, M. J.** Sensory, motor, and autonomic dysfunction: The nervous system in familial dysautonomia. *Neurology*, 21(5): 486-494, 1971.

Autopsy of a 1-year-old child with familial dysautonomia (FD) revealed striking reductions (about 50%) in neurons of the autonomic ganglia and dorsal root ganglia, and in the tongue's sensory axons. The first-born child of Jewish parents (there was no family history of FD), S showed respiratory difficulties at birth and failed to thrive. At age 1, an episode of gagging and apnea culminated in death. Brain and spinal cord showed no great abnormality. The cerebellar foliate white matter revealed diffuse slight rarefaction and scattered foamy macrophages. Cervical sympathetic ganglia showed a few foci of acute inflammation, and there were neuromata of non-myelinated nerves in the cervical sympathetic chain. There was little evidence for complete denervation of any sensory or motor function. Neuron loss probably causes sensory-pathway and nerve-conduction impairment as well as many of the pharmacologic and metabolic aspects of the disease. (37 refs.) - *B. Berman.*

New York University Medical Center
New York, New York

2834 BAILEY, ROSS R. Congenital bowing of the long bones. *Lancet,* 2(7716): 157-158, 1971. (Letter)

The S was a mentally defective youth with several congenital anomalies, including considerable bowing of both femora and tibiae. He was the eighth of 10 children, 8 of whom were living and mentally normal. One was stillborn with a congenital cardiac lesion. One brother has a kyphosis but normally shaped legs. The maternal grandfather had marked thoracic kyphosis and deformed femora but was mentally normal. A paternal great uncle was mentally defective but with normal skeleton. Birth was normal, and the mother received no drugs or radiation during pregnancy. Although the infant had peculiarly shaped legs, no concern was felt until lack of normal development was noted at 9 mo. He cut 9 large irregularly shaped teeth at 12 mos, two of which were lost at 9 yrs. A second dentition never occurred. He sat up at 10 mos and crawled at a normal age, but was unable to stand up and walk until more than 3 yrs. At 9 yrs he was institutionalized as an imbecile. Examined at age 16-3/4, in addition to bowed legs and the dental anomaly, he was observed to have a high-arched palate, a pronounced alternating strabismus, defective ocular coordination, slight cardiomegaly, and a slight wedging of the seventh thoracic body consistent with Scheuermann's disease. At 17-1/2 yrs S contracted infectious hepatitis. At 19 yrs, he developed tuberculous pyrothorax and was found to have chronic active hepatitis. (1 ref.) - *S. M. Rucker.*

Charing Cross Hospital Medical School
London W.6, England

2835 SENIOR, BORIS. Impaired growth and onychodysplasia. *American Journal of Diseases of Children,* 122(1): 7-9, 1971.

The composite of physical features in 6 short children studied suggests a recognizable clinical syndrome. Shortness of stature and tiny dysplastic toenails on one or more toes were the major findings common to all. Other features present, although less consistently, were similarity of facial appearance (broad nose, prominent nares, wide mouth), abnormalities of the fifth fingers and toes, and a degree of intellectual retardation. Three Ss had esotropia and 2 had small, somewhat stubby fingers. The 6 Ss were White, of both sexes, born to healthy unrelated parents of normal height. There were 19 siblings, all normal. Five Ss were considered short at birth (length of the sixth could not be evaluated because of premature birth). Two had short limbs, but the others had normal body proportions. All were below the third percentile in height, but growth rates were parallel to the third percentile in 3 Ss and somewhat greater in the other 3 Ss. One S, not formally tested, was obviously of normal intelligence. The others had a moderate reduction in intelligence quotient. Those of school age attended regular schools, but had repeated grades in the past or were in special ungraded classes. Some variation in size and shape of toenails is common, but the minute, almost vestigial size and shape of the affected nails observed in these Ss is extremely rare. (4 refs.) - *S. M. Rucker.*

171 Harrison Avenue
Boston, Massachusetts 02111

2836 LOWRY, R. B.; MacLEAN, R.; McLEAN, D. M.; & TISCHLER, B. Cataracts, microcephaly, kyphosis, and limited joint movement in two siblings: A new syndrome. *Journal of Pediatrics,* 79(2): 282-284, 1971.

Two siblings (a boy, and a girl 18 months younger) present with similar combinations of congenital malformations which do not correspond with previously described syndromes. This new syndrome involves the central nervous system, the eyes, and the musculoskeletal system: each child exhibits microcephaly, severe MR, cataracts, kyphosis, and limitation of joint movements. Results of chromosome studies on both parents were normal, and parents are otherwise healthy and normal. Therefore, this is probably a new autosomal recessive gene syndrome, and the parents have been counselled that risk of recurrence for subsequent pregnancies is as high as 25%. (6 refs.) - *K. L. Woodworth.*

715 W. 12th Avenue
Vancouver 9, British Columbia, Canada

2837 SEEMANOVA, EVA. A study of children of incestuous matings. *Human Heredity,* 21(2): 108-128, 1971.

A series of 161 children resulting from incestuous matings was compared with a control sample of 95 children derived from matings between the same mothers and unrelated partners. The series consisted of 88 children from father-daughter unions, 72 from brother-sister unions, and 1 from a mother-son union. Although no information concerning early fetal losses was available, the obtained data showed an unmistakable effect of inbreeding on infant mortality, congenital malformations, and intelligence level. However, the series was relatively small, and a direct comparison with the data from legally permitted consanguineous unions was not possible. The parents of the children of incest are not a representative sample of the general population, and specific adverse factors tend to accompany a pregnancy of incestuous origin in a marked way. Moreover, the sample of incestuous parents was not homogenous even with respect to the age of the mothers or their mental standard. Twenty of the 141 mothers were MR; of the 138 fathers, 8 were MR. (3 refs.) - *B. J. Grylack.*

Ustav vyzkumu vyvoje ditete V uvalu 84
Prague 5-Motol, Czechosolvakia

2838 **SAHAR, A.; HOCHWALD, G. M.; & RANSOHOFF, J.** Cerebrospinal fluid turnover in experimental hydrocephalic dogs. *Neurology*, 21(3): 218-224, 1971.

Cerebrospinal fluid (CSF) dynamics in experimentally produced hydrocephalus were examined in 20 dogs. The rates of formation and absorption of CSF were studied by lateral ventricle to lateral ventricle perfusion. The opening pressure in the 20 hydrocephalic animals was a mean of 10.5 cm H_2O and 7 cm H_2O in 7 normal dogs. The mean rate of CSF secretion was 0.033 ml/minute in the hydrocephalic dogs as compared to 0.044 ml/minute in the normal dogs. The absorption rates of CSF were 0.0170 + 0.000421 P ml/minute in 16 hydrocephalic dogs and 0.0390 + 0.000627 P ml/minute in 7 normal dogs. Absorption of CSF was found to be dependent upon perfusion pressure within the range of 10 to 40 cm H_2O. The results which indicated a diminished rate of formation of CSF in the hydrocephalic dogs were similar to those obtained in previous experiments. The moderate degree of ventricular enlargement noted in this study and the relatively low pressure recorded suggested the possibility that a state of "equilibrium" existed in these animals, an equilibrium between formation and absorption of CSF under even such low pressure. It is therefore possible that perfusion of CSF space does not accurately reflect the rate of formation and absorption of the fluid in the intact animal. (20 refs.) - *B. J. Grylack.*

New York University Medical Center
New York, New York 10016

2839 **LEVIN, VICTOR A.; MILHORAT, THOMAS H.; FENSTERMACHER, JOSEPH D.; HAMMOCK, MARY K.; & RALL, DAVID P.** Physiological studies on the development of obstructive hydrocephalus in the monkey. *Neurology*, 21(3): 238-246, 1971.

Acute obstructive hydrocephalus was induced in 8 rhesus monkeys through surgical insertion of an inflatable balloon into the fourth ventricle of the anesthetized animals. The 8 hydrocephalic monkeys were then compared to 5 normal monkeys without hydrocephalus but with ventricular balloons in order to determine the permeability characteristics of the brain capillaries and parenchyma, to establish the size and compartmentalization of the enlarged "spaces" seen in light and electron micrographs of hydrocephalic brains, and to evaluate the cerebrospinal fluid (CSF) production rates in hydrocephalic monkeys. The effective CSF secretion rate in the normal and hydrocephalic monkeys was 14.8±4.3 (SD) and 12.8±3.2 (SD) μl per minute, respectively. These values are not statistically different ($0.4>p>0.3$). Experimentally determined distribution spaces for ^{14}C-sucrose, ^{3}H-mannitol, and ^{24}Na in normal and hydrocephalic caudate nucleus were similar, while the periventricular white matter spaces were significantly different. The transcapillary exchange of ^{24}Na for both white and gray matter was greater in the hydrocephalic animals. The permeability of brain capillaries appears to be altered by acute obstructive hydrocephalus. (21 refs.) - *B. J. Grylack.*

Massachusetts General Hospital
Boston, Massachusetts

2840 **HECK, ALBERT F.; HAMEROFF, STEVEN B.; & HORNICK, RICHARD B.**

Chronic Listeria monocytogenes meningitis and normotensive hydrocephalus. Case report and review. *Neurology*, 21(3): 263-270, 1971.

A case was reported involving a 65-year-old man with normotensive communicating hydrocephalus presenting as dementia and associated with chronic Listeria monocytogenes meningitis. This hydrocephalus was demonstrated by ventriculography, pneumoencephalography, Pantopaque ventriculography, and radioisotopic cisternography. Antibiotic treatment and a right ventriculoatrial shunting procedure utilizing a Pudentz valve and tubing helped to produce a marked improvement in the dementia and, thus, to allow the man to function adequately at skilled labor, outside the hospital. Chronic Listeria meningitis has rarely been reported. The dramatic, rapid improvement observed in the clinical status of this patient following ventriculoatrial shunting is characteristic, regardless of the etiology of the hydrocephalus, and occurs so frequently as to be considered a criterion for diagnosis of normotensive hydrocephalus by some authors. (27 refs.) - *B. J. Grylack.*

University of Maryland School of Medicine
Baltimore, Maryland 21201

2841 HALL, BRYAN D. Preponderance of Bartter syndrome among Blacks. *New England Journal of Medicine*, 285(10): 581, 1971. (Letter)

There may be an increased frequency of Bartter's syndrome (hypokalemic alkalosis, hyperaldosteronism, juxtaglomerular hyperplasia, MR, and short stature) among Blacks, based on cases reported in the American literature. In the 17 reported cases which mention race, 11 Black families and 4 Caucasian families are represented. Unpublished information on cases in 3 additional Black families brings the total to a frequency of 76.4% (13 families) Black to 23.4% (4 families) Caucasian, respectively. Since the number of patients is not yet adequate to permit conclusions, other American physicians with unreported cases should provide data on the race of their patients to determine if the Black preponderance is real. (2 refs.) - *K. L. Woodworth.*

No address

2842 GILLES, F. H.; & ROCKETT, F. X. Infantile hydrocephalus: Retrocerebellar "arachnoidal" cyst. *Journal of Pediatrics*, 79(3): 436-443, 1971.

Benign retrocerebellar cysts can cause hydrocephalus by obstructing the flow of cerebrospinal fluid from the fourth ventricle. After adequate demonstration by air study, the cyst wall may be removed surgically, providing relief of the hydrocephalus without a shunt in many cases. The benign retrocerebellar cyst can be differentiated from the Dandy-Walker malformation by ventriculographic and morphologic criteria. Presence of cuboidal epithelium and/or astroglia in the cyst wall suggests that the cysts may be remnants of a persistent Blacke's pouch (persistence of a diverticulum of the roof of the fourth ventricle, which failed to involute during development). In 11 children with retrocerebellar cysts and hydrocephalus, surgical removal of the cyst wall was successful in all cases, although 6 patients subsequently required shunts. Of the 5 without shunts, 4 were normal 1-9 years after the operative procedure. Four of the shunt-requiring patients were well 1-3 years after the shunting procedures. The remaining 2 died, 1 from infection 1 year after operation and 1 from infection and cerebral hemorrhage 3 years after operation. (29 refs.) - *K. L. Woodworth.*

Children's Hospital Medical Center
Houston, Massachusetts 02115

2843 RUVALCABA, R. H. A.; REICHERT, A.; & SMITH, D. W. A new familial syndrome with osseous dysplasia and mental deficiency. *Journal of Pediatrics*, 79(3): 450-455, 1971.

Two Caucasian brothers represent a previously unrecognized dysmorphic syndrome characterized by MR, short stature with delayed adolescence, microcephaly, peculiar facies, hypoplastic genitals and skin lesions, and skeletal deformities (short metatarsals and metacarpals, small phalanges with tufting, epiphysitis of the spine). Two maternal female cousins also have several of these unusual features. The mechanism of inheritance is undetermined. However, one possibility is X-linked semidominant inheritance, the expression in the female being predominantly dependent on the allele of the mutant X-linked gene. The mutant X-linked gene would be present in the hemi-

zygous fully expressed state in the boys. (7 refs.) - *K. L. Woodworth.*

Rainer School
Buckley, Washington 98321

2844 **DOUGALL, A. J.** Central venous pressure in infants. *Lancet*, 2(7719): 315, 1971. (Letter)

During the operation of insertion of a ventriculoatrial drain with Holter valve for hydrocephalus, the central venous pressure of 16 infants (ages 10 days to 3 months) was recorded. Maximum central venous pressure was +11c, the minimum -1.5cm, and the average +4.6cm water pressure. Maximum swing with respiration was 3.5cm, the minimum 0.3cm, and the average 1.2cm. There are few records of pressure in normal children, and a single central venous pressure measurement cannot be considered abnormal unless it is outside a wide range. (2 refs.) - *K. L. Woodworth.*

Hospital for Sick Children
London WCIN 3JH, England

2845 **MILHORAT, THOMAS HERRICK.** *Hydrocephalus and the Cerebrospinal Fluid.* Baltimore, Maryland, Williams & Wilkins, 1972, 237 p. (Price unknown)

With the present monograph, an attempt has been made to bring together some of the more important facts about hydrocephalus, with particular emphasis on its causes, effects, diagnosis, and treatment. The volume is intended primarily for clinicians, although the biological aspects of the disorder are stressed throughout. Extensive bibliographies follow each chapter. - *B. J. Grylack.*

CONTENTS: Cerebrospinal Fluid Physiology; Pathogenesis of Hydrocephalus; Pathology of Hydrocephalus; Pathophysiology of Hydrocephalus; Diagnosis of Hydrocephalus; Treatment of Hydrocephalus.

Children's Hospital National Medical Center
Washington, D. C.

2846 **PRYOR, HELEN B.** A case of Waardenburg's syndrome. *American Journal of Diseases of Children*, 122(2): 177-178, 1971.

A case of Waardenburg's syndrome was presented in order to provide a comparison between this disorder and true hypertelorism in girls of approximately the same age. The head measurements of the 3-year, 7-month-old girl with Waardenburg's syndrome were 47.1 cm circumference, 13.2 cm breadth, and 16.0 cm length, while the child with hypertelorism had measurements of 47.2, 13.2, and 15.4 cm, respectively. Facial measurements in Waardenburg's syndrome were 8.7 cm height, bizygomatic 9.5 cm, and bigonial 7.6 cm, compared with 7.8, 11.4, and 8.8 cm, respectively, in hypertelorism. Finally, the ophthalmological measurements in the syndrome were interpupillary distance of 4.80 cm, inter-inner canthus 3.6 cm, and inter-outer canthus 7.8 cm, as opposed to 6.85, 4.2, and 9.5 cm, respectively, in the other child. The girl with Waardenburg's syndrome also had albinism of eyebrow and eyelashes, blepharophimosis mimicking hypertelorism, posterior cleft palate, lateral displacement of inner canthi, prominent root of nose, mild eyebrow hypertrichosis, and MR. (8 refs.) - *B. J. Grylack.*

Stanford University Medical Center
Stanford, California 94305

2847 **COFFIN, GRANGE S.; & SIRIS, EVELYN.** Soft hand syndrome. *American Journal of Diseases of Children*, 122(2): 181, 1971. (Letter)

Two new cases of the soft hand syndrome have been described, and visits to 1 kinship have yielded a family tree of 4 generations of mother-to-child transmission. The diagnosis is begun by shaking the hand of the patient, which seems very flabby and jointless. Tapered fingers, wide-set eyes, short stature, and retarded or low normal intelligence are also characteristic. The roentgenologic features of the hand, especially the "tufted" terminal phalanges, may be almost pathognomonic. (1 ref.) - *B. J. Grylack.*

2848 **HOLMES, LEWIS B.** Norrie's disease: An X-linked syndrome of retinal malformation, mental retardation, and deafness. *Journal of Pediatrics*, 79(1): 89-92, 1971.

Studies were performed on an American family in which 6 males in 4 generations were affected

with Norrie's disease, an X-linked recessive syndrome characterized by retinal malformation, MR, and progressive deafness. As often occurs in patients with Norrie's disease, infants in this family were diagnosed as having retrolental fibroplasia, even though they had normal birth weights and no exposure to an increased oxygen concentration during the first days of life. The primary ocular defect is severe hypoplasia of the inner retinal layer. The shrinkage of the globe and cataracts and hyperplasia of the retinal, ciliary, and iris pigment epithelium are secondary changes. In addition to retrolental fibroplasia, the more common diseases of retinoblastoma and intrauterine infections, including congenital toxoplasmosis, rubella and herpes virus, should be considered before the clinical diagnosis of Norrie's disease is made in an infant or young child. (10 refs.) - *B. J. Grylack.*

Massachusetts General Hospital
Boston, Massachusetts

2849 SAKATI, NADIA; NYHAN, WILLIAM L.; & TISDALE, WILLIAM K. A new syndrome with acrocephalopolysyndactyly, cardiac disease, and distinctive defects of the ear, skin, and lower limbs. *Journal of Pediatrics*, 79(1): 104-109, 1971.

A patient with a new syndrome of acrocephalopolysyndactyly has been reported. The cardinal clinical features of this 8-year-old male were cranial synostosis, acrocephaly, an unusual facies, brachydactyly and polydactyly of the fingers, polydactyly and syndactyly of the toes, bowed femurs, hypoplastic tibias, posterior displacement of the fibulas on the femurs, and linear submental scars resembling clefts. The patient also had congenital heart disease. When studied at 8 years of age, he had never been operated upon for craniosynostosis, but signs of chronic increased intracranial pressure were mild, and his intelligence appeared to be within normal limits. He had learned to walk with crutches in spite of leg malformations and was attending school. Although the condition described represents a distinct syndrome, many of the clinical features of the patient have been seen in other syndromes. Embryologically, this syndrome appears to be related to Apert's syndrome and the Laurence-Moon-Bardet-Biedl syndrome as well as to other acrocephalopolysyndactylies. (15 refs.) - *B. J. Grylack.*

University of California at San Diego
Department of Pediatrics
La Jolla, California

2850 STARK, GORDON D. Neonatal assessment of the child with a myelomeningocele. *Archives of Disease in Childhood*, 46(248): 539-548, 1971.

An outline for neonatal assessment of the child with a myelomeningocele has been presented by a hospital which admits 35 new cases of myelomeningocele yearly. The initial assessment should be carried out by a pediatric neurologist, who is joined later by surgical and orthopedic staff. Particular attention is paid to the general state of the infant, which can have a profound influence on the neurological findings and fitness for operation. Close examination of the spinal lesion is important. Neurological evaluation includes cerebral function, spinal cord function, and lower limb deformity. Special investigations consist of bacteriological swabs from the myelomeningocele and umbilicus, photography of the spinal lesion and lower limbs, radiological examination of the skull, hips, and spine, concentric needle electrode electromyography, air ventriculography, and urinary tract evaluation. Neonatal assessment is a guide to immediate medical and surgical treatment and lays the foundation for planning of future management. (34 refs.) - *B. J. Grylack.*

University of Edinburgh
Edinburgh EH9 1UW, Scotland

2851 BAILEY, IAN C. Double meningocele. *Archives of Disease in Childhood*, 46(248): 549-550, 1971.

An unusual case of a newborn girl with 2 distinct meningoceles with a normal intervening spinal canal has been observed. Both the thoracic and lumbar lesions received successful surgical therapy. There was no associated hydrocephalus or apparent neurological defect on examination at 9 months of age. Double meningoceles reported previously have been associated with

hydrocephalus and/or Arnold-Chiari malformation. (4 refs.) - *B. J. Grylack.*

Mulago Hospital
Kampala, Uganda, East Africa

2852 **JUDGE, C.; & CHAKANOVSKIS, JOHANNA E.** The Hallermann-Streiff syndrome. *Journal of Mental Deficiency Research*, 15(Part 2): 115-120, 1971.

A mentally deficient 12-year-old boy was diagnosed as having the Hallermann-Streiff syndrome on the basis of proportionate dwarfism (height 128 cm and weight 27.3 kg), malformed facies with mandibular and maxillary hypoplasia, markedly beaked nose, small mouth, microophthalmia, microbulbus, microcornea, aphakia, and partial anodontia with an absence of the upper lateral incisors, cuspids, bicuspids (first and second), and the lower bicuspids (first and second). Psychological examination showed the propositus to be grossly inferior for his age. Complete hematological tests, electroencephalography, and chromosomal and dermatoglyphic analysis revealed no abnormalities. The case reported here is atypical only in that aphakia has not been described previously. A review of the literature indicates that intelligence is not affected in some cases of the Hallermann-Streiff syndrome and that MR may be only mild in some individuals and the result of a visual handicap. (25 refs.) - *B. J. Grylack.*

Children's Cottages
Kew, Victoria, Australia

2853 **NEVIN, N. C.; SCALLY, B. G.; KERNOHAN, D. C.; & DODGE, J. A.** Hereditary gingival fibromatosis. *Journal of Mental Deficiency Research*, 15(Part 2): 130-135, 1971.

Gingival fibromatosis, epilepsy, MR, and bilateral camptodactyly were observed in a brother and sister. Gingival fibromatosis usually appears during and after eruption of the permanent teeth with a gradual enlargement of the gingival mucosa, but, as in these 2 Ss, it may occasionally present shortly after birth. Hypertrichosis is the anomaly associated most frequently with gingival fibromatosis. Although the Ss showed no hirsutism, this feature may not yet have developed. MR and epilepsy may also be associated anomalies. The 14-year-old boy has been a patient in a hospital for MRs for the past 10 years, and his youngest sister, now almost 7 years old, attends a school for MR. Review of 3 generations of the family did not reveal any evidence of manifestations of the disorder in relatives of the Ss. Although most cases of gingival fibromatosis are inherited in an autosomal dominant manner, autosomal recessive inheritance appears to be the most feasible form of transmission in this family and has been reported in other cases as well. (13 refs.) - *B. J. Grylack.*

The Queen's University
Belfast, Northern Ireland

2854 **STARK, GORDON.** Prediction of urinary continence in myelomeningocele. *Developmental Medicine and Child Neurology*, 13(3): 388-389, 1971.

Techniques which could predict bladder function and urinary continence in neonates with myelomeningocele would be extremely useful. Clinically, the vertebral level of the lesion is a poor predictor but neurological integrity of muscles innervated from S2-4 shows good correlation with detrusor activity. Partial continence defined as dry periods for more than one hour is a better prognostic sign than constant dribbling. Direct cystometry to rule out abnormal reflex activity and inert bladders is of great value in augmenting the clinical evaluation. (12 refs.) - *E. L. Rowan.*

Royal Hospital for Sick Children
Edinburgh, Scotland

2855 **LORBER, J.** Results of treatment of myelomeningocele. *Developmental Medicine and Child Neurology*, 13(3): 279-303, 1971.

Extensive followup of 524 unselected cases of myelomeningocele operated on shortly after birth suggests that criteria for selection for operation can be made on the basis of sequelae. The frequency of death and the extent of severe physical and mental handicap appeared independent of operative intervention in many cases. At the time of birth, evidence of severe paralysis (no

voluntary muscle activity in lower limbs or loss of innervation below L2-L3), head circumference at or over the 90th percentile, kyphosis severe enough to require osteotomy or associated gross congenital anomalies precluded normal development. No case with ventriculitis in the newborn period developed normally. If children meeting these criteria are denied massive operative intervention, then more effort might be directed toward those who could truly benefit from treatment and the children in question might be spared massive physical and psychic trauma. (15 refs.) - *E. L. Rowan.*

University of Sheffield Department of Child Health
Sheffield S10 2TH, England

2856 MACKEITH, R. C. A new look at spina bifida aperta. *Developmental Medicine and Child Neurology*, 13(3): 277-278, 1971. (Editorial)

Routine immediate operative closure of all myeloceles and meningomyeloceles at birth has resulted in the necessity for extensive followup procedures on the brain, bladder and legs and in psychosocial intervention in the lives of these children. Results have not been particularly heartening in some cases. The physician plays a key role in predicting the outcome of such heroic treatment and should have the option of suggesting that it be withheld. Many moral and ethical issues must be resolved if this is to become an acceptable alternative and a great deal of courage will be necessary in order to follow through. (4 refs.) - *E. L. Rowan.*

2857 Weighing the adrenals in hyaline membrane disease. *Medical World News*, 12(17): 21, 1971.

Examination of 387 newborn humans who lived less than 72 hours revealed that the adrenal glands of the 220 with hyaline membrane disease (HMD) were 19% lighter than those of babies who died from other causes. At the highest gestational ages, weight differences were most striking in that the adrenal glands of full term HMD babies weighed only 63% of normal. The adrenal glands of children exposed to antenatal bacterial infections were 16% heavier than those of noninfected babies. Infection appears to have a protective effect against HMD. Examination of 14 anencephalic neonates revealed that they had only 45% as many osmiophilic granules per volume in their lung tissues as nonanencephalic neonates. Data from experiments with fetal animals indicate that hormones may someday be successfully used to treat or prevent HMD. Hormones have been used in animals to stimulate early surfactant production *in utero* and to accelerate fetal lung maturation. The therapeutic implications of these findings for humans are unclear. (No refs.) - *J. K. Wyatt.*

2858 MURPHY, JOHN F.; & MULCAHY, RISTEARD. The effect of age, parity, and cigarette smoking on baby weight. *American Journal of Obstetrics and Gynecology*, 3(1): 22-25, 1971.

Investigation of 10,692 mothers who had live births, stillbirths and neonatal deaths, revealed that while age had no significant effect, cigarette smoking was related to a significant reduction in baby weight. Average baby weight is related to the consumption of cigarettes of up to 12 per day, the correlation coefficient being 0.77. Evidence suggests that retardation of fetal growth occurs early in pregnancy, possibly due to increased levels of carboxyhemoglobin in the fetal blood. (13 refs.) - *R. Seevaratnam.*

Birmingham and Midland Hospital for Women
Birmingham 11, England

2859 IYENGAR, LEELA. Folic acid requirements of Indian pregnant women. *American Journal of Obstetrics and Gynecology*, 3(1): 13-16, 1971.

Folic acid deficiency in pregnancy, especially in the last trimester, is very high in low income group Indian women. In investigation of this deficiency, 100, 200, 300 μg of folic acid with 60 mg of elemental iron was given daily to 200 women between 20-24 weeks of pregnancy. Controls received only 60 mg of elemental iron as ferrous fumarate. Hemoglobin was estimated as oxyhemoglobin and packed cell volume by microhematocrit. Whole blood folate was microbiologically assayed using *L.casei*. Six weeks after delivery, red folate cell levels both in controls and the 100 μg group were lower than at term

while they were similar in the 200 μg group and higher in the 300 μg group. Birth weights of infants in the 200 and 300 μg group were higher, as also was the red folate level in infants of the 300 μg group. Although the folic acid requirement is concluded to be 300 μg per day it was recommended that 500 μg of folic acid and 60 mg of iron be administered daily in the last 100 days of pregnancy. (9 refs.) - *R. Seevaratnam.*

Jamai-Osmania
Hyderabad, India

2860 D.N.A. synthesis and birth weight. *Lancet*, 2(7729): 856-857, 1971.

Infants whose low birth weight is appropriate for their shortened gestation period and those whose weight is disproportionately low (light for date - LFD.) differ in their liability to postnatal metabolic hazards and in their prognosis for long-term growth and development. Viral infections and chromosome aberrations are associated with intrauterine growth retardation. DNA synthesis and cell division are delayed in G-trisomy cells, and virus infections inhibit mitosis and increase the number of chromosome breaks. It is likely that these influences in the active stage of cell multiplication during early pregnancy lead to an irrecoverable reduction of cell number in the human fetus, thus explaining the persistence of growth retardation. Examination of peripheral blood culture of children with severe protein-calorie malnutrition revealed structural chromosome aberrations, due either to protein deficiency or to the greater susceptibility of such children to viruses, radiation, or drugs. In the face of evidence of impairment of transport of aminoacids to the fetus in hypertensive women, similar investigations in LFD infants will be interesting. (13 refs.) - *R. Seevaratnam.*

2861 **GAL, ISABEL.** Mongoloid features associated with spina bifida, a preliminary communication. *Developmental Medicine and Child Neurology*, 13(3): 380-383, 1971.

Eleven of 110 cases of spina bifida also showed prominent mongoloid features in general facial appearance and single palmar creases. Karyotypes were performed on 10 and none had Down's syndrome. As a group the infants with mongoloid features had higher mortality and lower intelligence than the rest of the study group. A larger study is necessary in order to assess the importance of these findings. (6 refs.) - *E. L. Rowan.*

Queen Charlotte's Maternity Hospital
London W. 6, England

2862 **GLASS, LEONARD; KOLKO, NORMA; & EVANS, HUGH.** Factors influencing predisposition to serious illness in low birth weight infants. *Pediatrics*, 48(3): 368-371, 1971.

A study was done to identify 4 factors associated with high rates of rehospitalization among low birth weight infants following discharge from a nursery. Ninety-nine infants were evaluated retrospectively and 79 infants prospectively, with particular attention paid to the failure of the mother to receive prenatal care, absence of the father from the home, receipt of public assistance, and presence of other children in the home. These 4 sociomedical factors were demonstrated to predispose to very high rates of serious illness, primarily infections, and poor follow-up medical care in low birth weight infants during the first 9 months of life. A prognostic index devised from the retrospective phase of the study proved significantly successful in the prospective phase. A special worksheet based in part upon the prognostic index and summarizing potential risk, visiting nurse reports, and clinic attendance was implemented by social workers in contact with families of low birth weight infants. (10 refs.) - *B. J. Grylack.*

532 Lenox Avenue
New York, New York 10037

2863 **WOLSTENHOLME, G. E. W.; & O'CONNOR, MAEVE, eds.** *Foetal Autonomy: A Ciba Foundation Symposium.* London, J. & A. Churchill, 1969, 326 p. $10.50.

In the account of the Ciba Conference on Foetal Autonomy, approximately 25 professionals speak on at least 1 topic. The subjects presented include fetal metabolism, fetal circulation, and

the role of the fetus in the initiation of parturition in the ewe. - *B. J. Grylack.*

2864 WAISMAN, HARRY A.; & KERR, GEORGE R., eds. *Fetal Growth and Development.* New York, McGraw-Hill, 1970, 320 p. $17.50.

This book is essentially a collection of 25 essays ranging beyond prenatal life into the aftereffects of disturbances of growth and development *in utero.* The presentations represent the proceedings of a conference sponsored by the University of Wisconsin School of Medicine and the Johnson & Johnson Institute for Pediatric Service. Topics include placental and fetal growth, monitoring of the intrauterine state, fetal diagnosis, determinants of fetal growth, abnormalities of fetal growth, and postnatal sequelae of intrauterine disease. - *B. J. Grylack.*

2865 STAVE, UWE, ed. *Physiology of the Perinatal Period: Functional and Biochemical Development in Mammals.* New York, Appleton-Century-Crofts, 1970, 1088 p. 2 volumes, $59.50 per set.

This 2-volume work contains studies by 41 contributors, 18 of whom represent countries other than those of North America. The books are subdivided into 35 chapters, the longer chapters being essentially monographs rather than essays. Topics presented include respiration and the respiratory system, the hemoglobins, heat production and temperature regulation, respiratory gas transport characteristics of blood and hemoglobin, neonatal hypoxia, metabolism. - *B. J. Grylack.*

2866 DUC, G. Assessment of hypoxia in the newborn. Suggestions for a practical approach. *Pediatrics,* 48(3): 469-481, 1971.

Measurement of oxygen partial pressure in arterial blood (Pao_2) is part of the routine management of cardiorespiratory problems in the newborn, the diagnosis of hypoxia usually being based upon that calculation. Other measurements used in the classification of neonatal hypoxia are the affinity of hemoglobin for oxygen (P_{50}), carbon dioxide partial pressure in arterial blood ($PaCO_2$), and oxygen saturation of hemoglobin. The 4 types of neonatal hypoxia which can be distinguished are hypoxia with hypoxemia due to a decrease in Pao_2 (A), hypoxia with hypoxemia due to a decrease in blood oxygen content without a decrease in Pao_2 (B), hypoxia with normoxemia due to a decrease in peripheral blood flow (C), and hypoxia with normoxemia due to an increase in P_{50} (D). Alveolar hypoventilation and right-to-left cardiac shunt are examples of A, anemia is an example of B, cardiac failure and shock are instances of C, and hyaline membrane disease represents the combination of different types of hypoxia. (22 refs.) - *B. J. Grylack.*

University Department of Pediatrics
and Obstetrics
8006 Zurich, Switzerland

2867 NELLHAUS, GERHARD. Head circumference in black and white and Indian (New Delhi) children. *Pediatrics,* 48(3): 494-495, 1971. (Letter)

Ethnic background plays little, if any, role in head circumference growth. Because of questions raised concerning single measurements of head circumference which fall between 2 and 3 standard deviations from the means, emphasis should be placed on the importance of serial measurements to determine head growth rate. Clinically, this is particularly important in determining abnormal deceleration or acceleration (progressive microcephaly or macrocephaly). (6 refs.) - *B. J. Grylack.*

University of Colorado Medical Center
Denver, Colorado 80220

2868 BRACKBILL, YVONNE. The role of the cortex in orienting: Orienting reflex in an anencephalic human infant. *Developmental Psychology,* 5(2): 195-201, 1971.

Habituation of the orienting reflex was studied in a male anencephalic infant. Experimentation dealt with the effects of stimulus intensity on probability and amplitude of the response, the possible occurrence of habituation in an anencephalic organism, the effects of interstimulus

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interval on habituation rate, and the influence of continuous stimulation on arousal level. Results showed that orienting reflex occurrence was a curvilinear function of stimulus intensity, the fewest responses occurring when low-moderate stimulus intensity values were employed. In comparison with data from normal infants, the responsiveness of the anencephalic S to stimulation was equal, but his ability to inhibit responses or to modulate their amplitude was impaired or absent. The cortex seems to be instrumental both in inhibiting response and in damping the amplitude of those responses that do occur. (8 refs.) - *B. J. Grylack.*

Georgetown University Hospital
Washington, D. C. 20007

2869 **GREEN, H. GORDON; GAREIS, FRANK J.; SHEPARD, THOMAS H.; & *KELLEY, VINCENT C.** Cretinism associated with maternal sodium iodide I 131 therapy during pregnancy. *American Journal of Diseases of Children,* 122(3): 247-249, 1971.

A case of cretinism following administration of sodium iodide I 131 to a pregnant woman with Grave's disease is described. Seven months prior to delivery, the mother was given I 131. A clinically normal infant was born, but at 4 years of age she was slow, inactive, tired easily and had coarse features. Her extremities were stubby, she did not speak or walk. Thyroid scan revealed near-absence of the right lobe of the gland. Levothyroxine sodium therapy was begun but the family left the area and the child was lost to follow-up. Congenital hypothyroidism may be a consequence of maternally administered radioactive iodine, the iodine having gross detrimental effects on the fetal thyroid. Hypothyroidism may be caused by ablation of the fetal thyroid by radioactive iodine; or the material may only partially injure it, leaving a remnant which functions for a variable period after birth. Radioactive iodine therapy is contraindicated in pregnant women and since pregnancy may not be suspected in many cases, other therapy for hyperthyroidism are to be preferred in women of childbearing age. If I 131 is given inadvertently in early pregnancy, termination of the pregnancy might be justified. If therapeutic abortion is not carried out, the infant should be closely watched, with long-term follow-up of growth and developmental progress. (11 refs.) - *T. C. Turner.*

*University of Washington School of Medicine
Seattle, Washington 98105

2870 **BLACKBURN, MICHAEL G.; & BELLIVEAU, ROBERT E.** Ellis-van Creveld syndrome. *American Journal of Diseases of Children,* 122(3): 267-270, 1971.

Another occurrence of the Ellis-van. Creveld syndrome is presented and association with previously undescribed anomalies is reported. Two cases are described: a female infant born to a normal mother had (hydrocephalus was discovered before birth) normal skin, nose, ears, eyes, mouth, liver and spleen, extra digits on both hands, partial syndactyly, acromelic shortening of the tubular bones, flared distal radial and proximal ulnar ends and distal femoral and tibial ends, with "trident" appearance of the acetabular roof. The mother's next child (female) had 6 digits on each hand with dysplastic nails and acromelic shortening of bones and flaring ends as in the first infant. Both had the same cardiac defect. These cases exemplify the typical clinical findings in Ellis-van Creveld syndrome, and exhibit other anomalies: congenital heart disease, and renal anomalies. Other anomalies which have been noted are neuroectodermal injury, absence of corpus callosum, hydrocephalus. An autosomal recessive pattern of inheritance of the Ellis-van Creveld syndrome has been established, but the manifestations of the syndrome may differ in siblings. The syndrome may also manifest itself in incomplete forms and isolated components of the syndrome may be present in patients' relatives. This partial manifestation should be considered in family genetic counseling. (19 refs.) - *T. C. Turner.*

333 Cedar Street
New Haven, Connecticut 06510

2871 **CHAKANOVSKIS, JOHANNA E.; & SUTHERLAND, G. R.** The Smith-Lemli-Opitz Syndrome in a profoundly retarded epileptic boy. *Journal of Mental Deficiency Research,* 15(3): 153-162, 1971.

A case of an 8-year-old PMR physically handicapped boy (with Smith-Lemli-Opitz syndrome, comprising a uniform pattern of multiple congenital anomalies of head, face, extremities and external genitalia) with seizures, intermittent hyperpnoea followed by apnoeic spells, cyanosis and ataxia is presented. The S came from a normal family with good health and had 2 normal sisters, but was premature, with extremely poor weight gain, head lag and respiratory distress. He could not speak, had evidence of plagiocephaly, knobby knees and ankles, malpositioned thumbs and toes, and a basic MA of 7 months. The S has most common features of the Smith-Lemli-Opitz syndrome with the exception of syndactyly of the second and third toes. It is suggested that this condition is due to a cytogenetically undetectable chromosome abnormality rather than to an autosomal recessive genetic disease (19 refs.) - *T. C. Turner.*

Janefield Training Center
P. O. Bundoona 3083
Victoria, Australia

2872 **FARNSWORTH, P. B.; & PACIK, P. T.** Glossoptotic hypoxia and micrognathia—the Pierre Robin syndrome reviewed. *Clinical Pediatrics,* 10(10): 600-606, 1971.

The Pierre Robin syndrome, which is often a newborn emergency because of glossoptotic hypoxia, is confusing to clinicians because of the lack of uniformity regarding the definition of the syndrome. Glossoptosis and micrognathia are always included, but the presence or absence of a cleft palate or other anomaly is open to argument. No definite etiology has yet been found to explain the syndrome, although theories include congenital syphilis or TB, infection, intra-uterine pressure or alteration of an endogenous substance during embryogenesis, resulting in mandibular malformation and ptosis, as possible causes for the condition. Ocular and bony defects and congenital heart disease are some of the many congenital anomalies which may be associated with micrognathia and glossoptosis. The initial management of a newborn with this syndrome is the permanent correction of glossoptosis obstructing the respiratory passage; then, management of associated anomalies can be carried out. Other methods of intervention include: placing the newborn infant in the prone position to keep the tongue forward, using an acrylic obturator, applying suture-traction to the tongue, performing tracheostomy or temporary deep suture glossopexy, surgical anterior fixation of the tongue. Feeding can be done by "orthostatic feeding" or a temporary gastrostomy. Ultimate mandibular development cannot be predicted, however. (37 refs.) - *T. C. Turner.*

Grasslands Hospital Division of Pediatrics
Valhalla, New York 10595

2873 **SIEBEN, ROBERT L.; HAMIDA, MONGI BEN; & SHULMAN, KENNETH.** Multiple cranial nerve deficits associated with the Arnold-Chiari malformation. *Neurology,* 21(7): 673-681, 1971.

Of 15 infants with myelomeningocele (MM) and the Arnold-Chiari malformation (ACM), 4 showed clinical signs of multiple deficits of the 6th, 7th, 9th, 10th, 11th, and 12th cranial nerves. Laryngeal stridor resulting from abductor paralysis of the vocal cords has been found associated with MM and ACM. One autopsy revealed nucleus ambiguus changes apparently secondary to cranial-nerve injury. Evidence shows that impaired functioning of other lower cranial nerves is not nearly so dramatic or life-threatening as when the vagus is involved. Laryngeal stridor and other aspects of lower cranial dysfunction are not apparently associated with other forms of hydrocephalus. The association may be due to herniation of posterior fossa structures into the cervical spinal cord. (21 refs.) - *B. Berman.*

Albert Einstein College of Medicine
New York, New York

2874 **BAUM, J. DAVID; & SEARLS, DONALD.** Head shape and size of newborn infants. *Developmental Medicine and Child Neurology,* 13(5): 572-575, 1971.

Measurements were made on 55 healthy newborn infants in order to consider the mathematical interrelationship among head measurements other than circumference and biparietal diameter, to determine whether the inclusion of more than 1 measurement of head size would provide an improved correlation with birthweight and gesta-

tional age, and to compare findings with those obtained on growing short-gestation infants. As a result of measurements of occipitofrontal circumference (OFC), anteroposterior (AP) and biparietal (BP) diameters, and cross-sectional area (A), equally close correlations were established for head size with gestational age and birthweight for infants appropriate in weight for their gestational age. From this study on heads of normal shape, formulas were established interrelating OFC, AP, and BP diameters, and A. When the size of a head of unusual shape is being considered, calculations from these measurements will yield the circumference of that A as if it were of normal configuration. (6 refs.) - *B. J. Grylack.*

Hammersmith Hospital
London W. 12, England

2875 **BAUM, J. DAVID; & SEARLS, DONALD.** Head shape and size of preterm low-birthweight infants. *Developmental Medicine and Child Neurology*, 13(5): 576-581, 1971.

Head circumference, shape, and cross-sectional area were measured in 14 preterm low-birthweight infants and in 45 young adult "ex-premature" patients in a study of the dynamics of head growth. All of the infants showed a pattern of progressive skull deformation with postnatal age. In the comparison of the 45 adults with normals, anteroposterior (AP) diameters were found to be significantly greater (X^2=11.7, $p<.05$) in the "ex-prematures," biparietal (BP) diameters were significantly smaller (X^2=51.5, $p<.001$), and occipitofrontal circumferences (OFCs) were significantly smaller (X^2=14.35, $p<.003$). Skull deformity of low-birthweight infants thus persisted into adult life. It appears that much of the molding found in preterm infants resolves before adulthood, but there is a persistence of the skull deformation acquired neonatally. (6 refs.) - *B. J. Grylack.*

Hammersmith Hospital
London W. 12, England

2876 **WHITE, JOHN J.** Membrane oxygenator can aid premature infants in respiratory distress. *Journal of the American Medical Association*, 215(11): 1742-1743, 1971.

A preliminary clinical trial indicates that prolonged respiratory support of premature infants in severe respiratory distress with a membrane oxygenator is practical and feasible. The spiral coil oxygenator provided excellent oxygenation, carbon dioxide removal, and good support in 3 infants for 3 to 10 days. The veno-venous bypass technique returned oxygenated blood via the umbilical cord, in effect mimicking fetal circulation and triggering a reflex which stopped ventilation. The technique holds promise for a variety of patients with acute but reversible respiratory failure. (No refs.) - *S. Glinsky.*

Johns Hopkins School of Medicine
Baltimore, Maryland

2877 **MORGAN, THOMAS E.** Pulmonary surfactant. *New England Journal of Medicine*, 284(21): 1185-1193, 1971.

Recently considerable advances have been made in knowledge of the pulmonary surfactant system. A deficiency of surfactant exists in the respiratory-distress syndrome of the newborn. The typical features of this syndrome include diffuse atelectasis, transudation of fluid, formation of hyaline membranes, labored breathing, retraction of the chest cage, and cyanosis. Recovery from the syndrome is connected with return of surface tension and saturated lecithin to normal levels. Other procedures and pathologic conditions which affect surfactant include pulmonary arterial occlusion, pulmonary edema, possible pulmonary alveolar proteinosis, air pollution, and certain surgical procedures. The beneficial effects of aerosolized surface-active agents seem questionable. (106 refs.) - *S. Glinsky.*

University of Washington School of Medicine
Seattle, Washington 98105

2878 **SOMMER, ANNEMARIE; CONTRAS, STELLA B.; CRAINEN, JO M.; & HOSIER, DON M.** A family study of the leopard syndrome. *American Journal of Diseases of Children*, 121(6): 520-523, 1971.

In a family consisting of a mother and 4 children, 3 of the children exhibited the findings of the recently described leopard syndrome. The

abnormalities found variously in the children included multiple lentigines, electrocardiographic conduction defects, ocular hypertelorism, and pulmonary stenosis. Performance in school ranged from normal to below average. The inheritance appears to be autosomal. (4 refs.) - *S. Glinsky.*

Ohio State University College of Medicine
Columbus, Ohio 43210

2879 **LOWRY, BRIAN; MILLER, JAMES R.; & FRASER, F. CLARKE.** A new dominant gene mental retardation syndrome. *American Journal of Diseases of Children,* 121(6): 496-500, 1971.

Many members of 1 family exhibit a new syndrome with MR and other characteristic features. The syndrome includes MR, small stature, retarded bone age, hypotonia, tapering fingers, characteristic facies of hypertelorism, upturned nares, and prominent frontal region. Arrested hydrocephalus possibly also exists. The evidence suggests that the cause of the syndrome is an autosomal dominant gene with variable expressivity. This family illustrates the importance of genetic counseling, which could have diagnosed MR much sooner. (7 refs.) - *S. Glinsky.*

University of British Columbia
Vancouver, British Columbia, Canada

2880 **COHEN, RICHARD L.** Pregnancy stress and maternal perceptions of infant endowment. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 313-320.

Stress incurred at any point during the adaptational process of pregnancy may seriously interfere with a favorable outcome of that process. Five cases illustrate this effect, which in many instances distorts the mother's capacity for objective perception of the infant's capabilities. Pregnancy stresses often are converted into fears about the infant's survival or about his state of physiological intactness. Little relationship seems to exist between the nature of the stress and the subsequent pattern of maternal fears. Recognition of these pathological child rearing attitudes as early as possible is important, because they may become strong influences in producing adverse developmental outcome in the child. (10 refs.) - *S. Glinsky.*

University of Pittsburgh School of Medicine
Pittsburgh, Pennsylvania

2881 **GLEICHERT, JAMES E.** Patterns in perinatal mortality. *American Journal of Obstetrics and Gynecology,* 107(5): 780-786, 1970.

The decline in the perinatal mortality rate in the United States has been slower since 1950; it is higher than in 6 Western European countries. A survey of a Dallas hospital's perinatal mortality rates between 1960 and 1968 encompassed 28,313 births and 689 perinatal deaths. Immaturity was the largest single factor and also was to be associated with all other categories in 66% of the deaths. A relationship seems to exist between the increasing number of deaths from respiratory distress in the immature neonate and the amount of sedation and type of anesthesia used during premature labor and delivery. (5 refs.) - *S. Glinsky.*

Methodist Hospital of Dallas
Dallas, Texas

MEDICAL ASPECTS — Etiologic Groupings
Gross brain disease (postnatal)

2882 **MONTPETIT, VITAL J. A.; ANDERMANN, FREDERICK; CARPENTER, STIRLING; FAWCETT, JOHN S.; ZBOROWSKA-SLUIS, DANUTA; & GIBERSON, HERBERT R.** Subacute necrotizing encephalomyelopathy. A review and a study of two families. *Brain,* 94(1): 1-30, 1971.

A study of two families in which more than one member was affected by subacute necrotizing encephalomyelopathy (SNE), a disease of unknown etiology, has provided additional support for the autosomal recessive mode of transmission postulated. In one family of 7 siblings, 3 died of SNE and 3 are suffering from the illness. In the second family of 4 sibs, 2 have died. SNE was confirmed histologically in 2 and 1 of the first and second families, respectively. Clinical observation in 53 reported cases has shown that the illness usually becomes manifest during the first 2 years of life; of this group, 39 patients died before the age of 4 years. A slight preponderance of affected males over females occurred in these cases. Comparison of the pathology of 50 cases of SNE with that reported for Wernicke's disease has uncovered both similarities and differences. Lactic acidosis is likely a secondary metabolic abnormality unrelated to the basic process; it may be related to metabolic acidosis of hyperventilation, since the reported elevation of blood lactate in 4 cases of SNE was not observed in the present study and in 2 other reported cases. The reported finding of an inhibitor in tissue fluid of TPP-ATP phosphotransferase requires additional study. (49 refs.) - *M. S. Fish.*

University of Ottawa
Ottawa, Canada

2883 **PINCUS, JONATHAN H.; COOPER, JACK R.; ITOKAWA, YOSHINORI; & GUMBINAS, MARIA.** Subacute necrotizing encephalomyelopathy. *Archives of Neurology*, 24(6): 511-517, 1971.

Initial success in the treatment of a case of subacute necrotizing encephalomyelopathy (SNE) (Leigh's disease) with thiamine and thiamine propyl disulfide suggests the possibility that these materials may be of use in the management of this disorder. The S was a 15-month-old male with progressive hypotonia, weakness, and lethargy who seemed to regress after the age of 14 mo. A female sib had developed similar symptoms at the age of 9 mo, and a second female sib lost appetite and weight and developed symptoms of failure to thrive at 10 mo. She died at 16 mo of respiratory distress. The dizygotic twin and an older sib of the S were normal. Examination of the urine of the S revealed marked inhibition of TPP-ATP phosphoryl transferase, and a course of oral thiamine was instituted, followed by marked improvement. During the course of treatment by administration via various routes of both thiamine and thiamine propyl disulfide, the maintenance of thiamine levels above 50 μg/100 ml in the cerebrospinal fluid became increasingly difficult in spite of blood levels of over 1,000 μg/100 ml; remissions and deterioration correlated with increased and decreased thiamine levels, respectively. An autopsy of the brain of the S after death showed that total thiamine content of brain tissue (2 μg/g) was slightly elevated over normal levels. Thiamine triphosphate was not present in the pons or cerebellum. (16 refs.) - *M. S. Fish.*

Yale University School of Medicine
New Haven, Connecticut 06510

2884 **AMMANN, ARTHUR J.; & HONG, RICHARD.** Autoimmune phenomena in ataxia telangiectasia. *Journal of Pediatrics*, 78(5): 821-826, 1971.

Comparison of clinical and laboratory features of patients with ataxia telangiectasia with those reportedly found in animals thymectomized at birth suggests that the thymus plays either a direct or indirect role in the development of autoimmune phenomena by predisposing to chronic infection. Quantitation of immunoglobulins and antibodies in 18 patients (ages 2 to 39 years) with ataxia telangiectasia and in 20 age-matched control Ss was performed. A pool of normal human serum was used to obtain proteins for immunization. Results of the tests showed that cellular immune deficiency (abnormality in response to phytohemagglutinin, in delayed hypersensitivity skin tests, and/or in total lymphocyte count) occurred in 12 patients. Deficiencies were noted in IgA and IgE for 9 and 11 patients, respectively, and 2 had deficiency in IgG, IgA, and IgE. In 6 patients abnormal kappa/lambda ratios were found. Tests for antibodies indicated the presence of those directed against IgG, IgA, and thyroglobulin in 5, 9, and 2 patients, respectively. Six patients had fluorescent antibodies: 3 against parietal cell; 2 (each) against smooth muscle, mitochondria, and striated muscle; and 1 (each) against bile canaliculi and tubular basement membrane. Five of 9 patients with IgA and/or IgE deficiency had autoimmune phenomena, as did 1 of 3 with abnormal cellular immunity but normal immuno-

globulins and 2 of 4 with both normal immunoglobulins and cellular immunity. The results suggest that the recurrent infections in ataxia telangiectasia are related to the presence of multiple immunologic defects which result from a thymic defect. (40 refs.) - *M. S. Fish.*

University of Wisconsin Medical Center
Department of Pediatrics
Madison, Wisconsin

2885 **GAJDUSEK, D. CARLETON; & GIBBS, CLARENCE J., JR.** Transmission of two subacute spongiform encephalopathies of man (kuru and Creutzfeldt-Jakob disease) to New World monkeys. *Nature*, 230(5296): 588-591, 1971.

More readily available New World monkeys may replace the chimpanzee in the laboratory study of two subacute degenerative diseases, kuru and Creutzfeldt-Jakob disease. Transmission of both diseases to the squirrel monkey and the spider monkey has been accomplished by intracerebral inoculation of 10% suspensions of human brain, followed by a 2-year incubation period. Intracerebral inoculation of human kuru brain suspension into the capuchin monkey causes the disease after 45 months. Experimental kuru in the spider monkey is transmissible to the squirrel monkey, and experimental Creutzfeldt-Jakob disease of the chimpanzee can be transmitted to the squirrel monkey and the woolly monkey. While both of these disorders have previously been passed successfully to the chimpanzee, the future use of 4 different species of smaller, less expensive, and more available New World monkeys will be of aid in these studies. Examination of 16 Old World species and 2 additional New World species has shown that, of the former group, the chimpanzee was the only species susceptible to these diseases, and the 2 other New World species were also resistant. (10 refs.) - *M. S. Fish.*

National Institute of Neurological
Diseases and Stroke
Bethesda, Maryland 20034

2886 **MOOSA, ALLIE; & DUBOWITZ, VICTOR.** Late infantile metachromatic leucodystrophy. *Archives of Disease in Childhood*, 46(247): 381-383, 1971.

A 3-year-old girl with metachromatic leukodystrophy was put on a vitamin A deficient diet and appeared to improve. After 6 weeks on the diet, which contained approximately 500-900 IU/day of vitamin A and supplementary vitamins C and D, she had improved considerably. During follow-up over a 2-year period, her condition remained static. She is unable to sit or stand, has complete aphasia, and cannot swallow solids. She has occasional extensor spasms, especially of the leg, and reflexes are all but absent. Nerve conduction velocity is very low. However, she is responsive and alert. (10 refs.) - *J. K. Wyatt.*

University of Sheffield
Sheffield, England

2887 **DESNICK, R. J.; DESNICK, S. J.; & DAWSON, GLYN.** Urinary screening of globoid-cell leukodystrophy. *New England Journal of Medicine*, 285(4): 239, 1971. (Letter)

In reply to the letter by Suzuki and Suzuki concerning the diagnosis of glycosphingolipidoses by urinary-sediment analysis, patient B.C. was identified as having a variant of Krabbe's leukodystrophy (KLD). Classification as such was based on demonstration of large amounts of galactosyl ceramide in red cells, bone marrow, plasma, urinary sediment, and cultured skin fibroblasts. We do not know whether globoid cells were present, since no brain biopsy was performed. In analyzing the urinary-sediment glycosphingolipids in a patient with juvenile KLD and another with classic KLD, no elevation of galactosyl ceramide was found in the urinary sediment, although glucosyl ceramide was significantly elevated in both cases. Without claiming that KLD can always be diagnosed by the presence of galactosyl ceramide in urinary sediment, the slightly increased levels of galactosyl and glucosyl ceramides in autopsy material do not preclude the possibility that the disease is present. (4 refs.) - *K. L. Woodworth.*

University of Minnesota
Minneapolis, Minnesota

2888 **SARSFIELD, J. K.** The syndrome of congenital cerebellar ataxia, aniridia and mental retardation. *Developmental Medicine*

and Child Neurology, 13(4): 508-511, 1971.

A syndrome of congenital cerebellar ataxia (a form of cerebral palsy), aniridia, and MR was observed in a white Caucasian male born in 1964, the second child of healthy, unrelated parents. Examination revealed slight hypotonia in all limbs, an ataxic and broad-based gait, indistinct and fragmented speech, loss of the pupillary portion of the irides and absence of the sphincter pupillae, and an IQ of 71 on the Merrill Palmer Scale. Chromosome studies showed a normal male karyotype. The patient has shown slow but steady improvement in balance and walking. Although reports on the familial incidence of congenital cerebellar ataxia are few, they have invariably suggested a recessive mode of inheritance. As for the aniridia, it was not due to dominant inheritance, and may represent a recessively inherited condition unrecognized until now. (9 refs.) - *K. L. Woodworth.*

University of Leeds
Department of Paediatrics and Child Health
Leeds LS1 3ET, England

2889 **SUZUKI, KUNIHIKO; & SUZUKI, YOSHIYUKI.** Urinary screening of globoid-cell leukodystrophy. *New England Journal of Medicine*, 285(4): 238-239, 1971. (Letter)

A patient referred to in an article by Desnick et al. was diagnosed as a case of Krabbe's globoid-cell leukodystrophy (GLD), but we do not agree with the diagnosis if the disease is defined as a genetic deficiency of galacto-cerebrosidose. In experience of 17 patients, all tissues, including cultured fibroblasts, of patients with GLD are always severely deficient in the activity of galactocerebrosidase, wherease B.C.'s enzyme activity was higher than in the other 4 patients and within the range of normal controls. In 5 patients, we observed with GLD there was no specific increase of galactocerebroside in the kidneys. Therefore, either the diagnosis of B.C. should be established by histology, or the excess urinary excretion of galactocerebroside should be confirmed on additional patients with genuine galactocerebrosidase deficiency, before the feasibility of the diagnosis of GLD by urinary-sediment analysis is established. (No refs.) - *K. L. Woodworth.*

University of Pennsylvania School of Medicine
Philadelphia, Pennsylvania

2890 **ANDREWS, JOHN M.; CANCILLA, PASQUALE A.; GRIPPO, JORGE; & MENKES, JOHN H.** Globoid cell leukodystrophy (Krabbe's disease): Morphological and biochemical studies. *Neurology*, 21(4): 337-352, 1971.

Clinical, routine neuropathologic, ultrastructural, chemical, and metabolic findings have been described in 3 cases of Krabbe's globoid cell leukodystrophy. The most prominent abnormality on light microscopy was the presence of single globoid cells and groups of them in the white matter, often adjacent to blood vessels, a typical finding in this disease. Ultrastructurally, both intracellular and extracellular crystalline-like inclusions were found. On the basis of ultrastructural data, it was speculated that the myelin loss evident in this disease was primarily, if not exclusively, due to axonal degeneration. However, this speculation was difficult to reconcile with the observation that axons may be relatively preserved in comparison to the degree of myelin loss, suggestive of primary demyelination, especially in its earlier stages. It also appears necessary to postulate chemical abnormalities other than the documented cerebrosidase deficiency in order to account for the initial axonal degeneration and the early loss of oligodendrocytes evident in this disease. (56 refs.) - *B. J. Grylack.*

The Center for the Health Sciences
Los Angeles, California 90024

2891 **JULIUS, R.; BUEHLER, B.; AYLSWORTH, A.; PETERY, L. ST.; RENNERT, O.; & GREER, M.** Diagnostic techniques in metachromatic leukodystrophy. *Neurology*, 21(1): 15-18, 1971.

A case of metachromatic leukodystrophy (MLD) in a girl aged 6 years 6 months was presented with documentation of the techniques which are currently available to diagnose the disease by enzymatic assay in urine, peripheral leukocytes, and fibroblast cultures and, for the first time, to define metachromatic granules in fibroblast cultures. Arylsulfatase A (ASA) is the specific

enzyme of relevance in MLD, and a marked diminution of that enzyme was determined by the appropriate techniques in this case. The specific use of cresyl violet in an aqueous medium yielding relatively specific histochemical data to establish the diagnosis of metachromatic leukodystrophy represents an initial success in that aspect of methodology. Therapeutic ideas related to MLD have included injection of ASA into a patient with MLD and the use of a vitamin A-deficient diet, neither of which has yet rendered any definite success. (17 refs.) - *B. J. Grylack.*

University of Florida College of Medicine
Gainesville, Florida 32601

2892 **ANDREWS, JOHN M.; SORENSON, VERNON; CANCILLA, PASQUALE A.; PRICE, HAROLD M.; & MENKES, JOHN H.** Late infantile neurovisceral storage disease with curvilinear bodies. *Neurology*, 21(3): 207-217, 1971.

Clinical and biochemical data on 3 patients with neurovisceral storage disease with curvilinear bodies (CLB), including 1 with associated intraneuronal zebra bodies (ZB), have been described. The patients were normal in 2 cases until 3 years and in 1 case until 3.5 years of age, after which neurological symptoms became apparent. On the basis of frontal lobe biopsy tissue analysis, the characteristic CLB were distinguished as well as the unusual ZB in 1 case. Biochemically, the outstanding results included an increase in ceramide trihexoside, reduction of hydroxy fatty acids and long-chain nonhydroxy fatty acids, and a normal ganglioside content in the tissue. Clinically, this disease cannot be separated readily from a variety of other childhood degenerative encephalopathies. Morphologically, although the CLB is a distinct feature, it seems that disease classification based upon a single morphological criterion is rarely more than a transient phase in the study of a group of related conditions which awaits further data. (35 refs.) - *B. J. Grylack.*

UCLA Center for the Health Sciences
Los Angeles, California 90024

2893 **HOFFMAN, PAUL M.; STUART, WILLIAM H.; EARLE, KENNETH M.; & BRODY, JACOB A.** Hereditary late-onset cerebellar degeneration. *Neurology*, 21(8): 771-777, 1971.

Clinical and pathological observations were recorded on a family in which hereditary late-onset cerebellar degeneration occurred in 19 members within 4 generations. The average age of onset was 54 years, and the disease was probably transmitted by autosomal dominance. Gait ataxia appeared first, with truncal instability frequently accompanying the ataxia or becoming evident 3 to 5 years later. Spastic scanning speech with a monotonous quality and little or no explosive tendency was also prominent but did not usually appear for several years following the onset of gait ataxia. Dementia was not a feature in most cases. Pathologically, the folia of the cerebellum were atrophied most severely in the superior and anterior portion of the vermis and hemispheres, the microscopic loss of Purkinje cells being most marked in these areas. Four cases in the fourth generation had an acute febrile illness preceding the ataxia. On the basis of this finding, it appeared possible that the febrile illness might have led to a more rapid deterioration of a susceptible cerebellum. (14 refs.) - *B. J. Grylack.*

National Institute of Neurological
Diseases and Stroke
Bethesda, Maryland 20014

2894 **O'REILLY, D. ELLIOTT.** The future of the cerebral palsied child. *Developmental Medicine and Child Neurology*, 13(5): 635-640, 1971.

On the basis of observations of 1,513 patients with cerebral palsy, the distribution of different types of handicap and the way in which these handicaps affects the patients' futures have been determined. Of the children studied, 60% were primarily spastic. Of the total, 34.4% were mentally normal, 25% were MR and had minimal physical handicap, and 25% had physical or mental handicaps of such magnitude as to require custodial care. The borderline and MR children with minimal physical problems required classes or schools for the MR. A large proportion of the patients who had average mentality and were self-sufficient was able to complete regular school and to participate in normal activities. The group of patients presenting a particularly difficult problem includes those with multiple handicaps which do not meet the requirements for care under any specific program. Only in some large metropolitan areas of the United States are the

needs of the handicapped met in any substantial way. (2 refs.) - *B. J. Grylack.*

St. Louis University School of Medicine
St. Louis, Missouri 63104

2895 **GARBER, NORMAN B.** Operant procedures to eliminate drooling behavior in a cerebral palsied adolescent. *Developmental Medicine and Child Neurology*, 13(5): 641-644, 1971.

A behavior modification program designed to reduce drooling was executed during speech therapy for articulatory disorders in a 14-year-old White male with choreoathetotic cerebral palsy and right hemiparesis. His speech was dysarthric, characterized by excessive nasality and omissions, distortions, and substitutions of consonant and vowel sounds. Constant drooling appeared to be due to the oral paresis. Positive reinforcement through implementation of a penny-reward system was the operant conditioning procedure chosen to increase nondrooling behavior. Results indicated a significance beyond the .01 level of confidence, showing a significant change in drooling behavior. This study demonstrates that behavior modification can be applied to problems of an organic nature, provided it is physiologically possible to change the behavior. (2 refs.) - *B. J. Grylack.*

University of Missouri
Columbia Speech & Hearing Clinic
Columbia, Missouri 65201

2896 **MACCULLOCH, M. J.; WILLIAMS, C.; & DAVIES, PAT.** Heart-rate variability in a group of cerebral palsied children. *Developmental Medicine and Child Neurology*, 13(5): 645-650, 1971.

Thirty psychometrically untestable cerebral palsied children with a mean CA of 8.25 years had on-line heart-rate recordings taken in order to test the hypothesis that nonpurposive movements in cerebral palsied children are accompanied by a raised heart-rate variability compared with nonathetoid cerebral palsied children. Of the 30 Ss, 20 were diagnosed as spastic and 10 as athetoid/dystonic. The groups consistently differed at a significant level on measures of heart-rate variability, with the athetoid/dystonic Ss showing a higher variability of heart rate than their spastic counterparts (coefficient of variation $t=3.06$, $p<0.01$; standard deviation $t=2.96$, $p<0.01$; and range $t=2.48$, $p<0.02$). The chance probability of the observed relationship between standard deviation and diagnostic category was less than 1 in 1,000 ($X^2=14.2$, $p<.001$). It has been hypothesized that the phenomenon of fluctuating tone, which is linked to arousal in athetoid cerebral palsied children, is related in part to the functions of the reticular formation, and specifically to an abnormally underdamped reticular formation. It may be predicted that athetoid children will show EEG abnormalities in arousal, and histological examination of the posterior brain stem at the level of the obex would be expected to show pathological changes related to neuronal dysfunction or loss. (16 refs.) - *B. J. Grylack.*

The University of Liverpool
Liverpool L69 3BX, England

2897 **ROBSON, PETER; & MAC KEITH, RONALD C.** Shufflers with spastic diplegic cerebral palsy: A confusing clinical picture. *Developmental Medicine and Child Neurology*, 13(5): 651-659, 1971.

Clinical findings were reported in the case of 9 children in whom shuffling behavior was complicated by an otherwise classical spastic diplegia. The spastic diplegic cerebral palsy did not emerge until the second or third year of life in these cases. At the time when the 5 earliest cases were seen, the features of the shuffling trait made the diagnosis uncertain, even after the appearance of the spasticity, and several alternative diagnoses had to be considered. Shortening of the hamstrings, brisk tendon jerks and persistent ankle clonus, and hypotonia present in the upper limbs and body axis but not in the lower limbs are early indications that a tendency to develop lower limb spasticity is present in any infant or child with hypotonia of the lower limbs. The majority of treatment regimens are based upon the assumption that all children will go through a stage of crawling. Therefore, it is possible that the shuffling child, who usually has a strong dislike of lying prone and typically does not crawl, might respond differently to such treatment regimens than the palsied child who crawls. (16 refs.) - *B. J. Grylack.*

Guy's Hospital
London S. E. 1, England

MEDICAL ASPECTS – Etiologic Groupings
Psycho-environmental

2898 AUG, ROBERT G.; & ABLES, BILLIE S. A clinician's guide to childhood psychosis. *Pediatrics*, 47(2): 327-338, 1971.

An attempt to resolve confusion about childhood psychosis has prompted a discussion of this wide array of conflicting descriptions and definitions from the point of view of the pediatrician. The best definition of childhood psychosis is the existence of a heterogeneous group of clinical syndromes, occurring between birth and age 11, which presents characteristic disturbances in: relationship with social environment; sense of personal identity; affect and its expression; use of speech for social communication; or total integration and organization of personality. The prevalence of psychoses in children is generally less common than in adults and is from 2.1 to 9/1000, depending on the criteria used. It is more common in boys than in girls (boy/girl ratio varies from 2:1 to 4:1). Labels applied to childhood psychosis include: early infantile autism, symbiotic psychosis, and childhood schizophrenia. Characteristic cardinal symptoms are: impairment of contact with the environment, of affect and its expression, of sense of personal identity, of speech, and unevenness of ego development. Nonspecific symptoms include: inability to grasp basic categories of thought; abnormalities in perception; disturbances of motility; impaired control of impulses; lack of defenses to handle anxiety; primitive thinking; obsessive insistence on sameness (this also occurs in some cases of MR and brain damage); and impairment of social skills. Childhood psychosis must be differentiated from: MR, behavior syndrome of minimal brain damage, and other conditions of delayed or absent speech, of "odd" or "different" behavior, and of hyperactivity and impulsivity. Opinions regarding the etiology of childhood psychosis vary from purely psychogenic to purely organic, and include various combinations of the two. Most etiologic research has been in the area of autism. (38 refs.) - *M. S. Fish.*

University of Kentucky Medical Center
Lexington, Kentucky

2899 SOLNIT, ALBERT J. A need for pediatric guidance. *Pediatrics*, 47(2): 325-326, 1971. (Commentary)

In the care of the psychotic (including autistic) child the pediatrician has a crucial and continuing role. He can provide vital guiding, interpreting, and protecting functions for these children and their parents if he properly utilizes current knowledge and is receptive to new programs and techniques. Early diagnosis and parental guidance and coordination of special services are essential for the care of these children. This approach is based on the assumptions that: biological characteristics exist which constitute vulnerability in these children and are expressed by how they relate to parents, to themselves, and to social experiences; childhood and adult psychoses differ, although important linkages may exist; when neurological deficiencies occur, the prognosis for psychotic children is poorer than when no organic deficiencies exist; special educational experiences are usually more effective for these children than psychotherapy or chemotherapy; biological research promises partial breakthrough in this area; and parents of these children require considerable support. (11 refs.) - *M. S. Fish.*

Yale University
New Haven, Connecticut

2900 WERSHOW, HAROLD J. Pathogenesis of urban slums. *Journal of the American Medical Association*, 215(12): 1959-1962, 1971.

Slums and poverty are self-perpetuating. The poor are brought up with models of failure. Inadequate medical care, inadequate stimulation, inadequate education, inadequate food, and inadequate jobs are the only available way of life. We must reorder priorities and spend vast sums of money on welfare, schools, job training, and adult education if the cycle is to stop. (3 refs.) - *E. L. Rowan.*

720 S. 20th St.
Birmingham, Alabama 35233

2901 **HOOD, CATRIONA.** Social and cultural factors in health of children of immigrants. *Archives of Disease in Childhood,* 46(247): 371-375, 1971.

A study of 100 1-year-old West Indian children and their families, with a follow-up study of 87 children at ages 2½ to 3 years, revealed that the children did not appear to be fulfilling their developmental potential. At age 1 year, the children had a higher incidence of respiratory disease, minor illnesses, and hospital stays than non-West Indian 1-year-olds. Performance on intelligence and social development scales at age 2½ to 3 years was poor in the verbal and manual dexterity areas. The children seemed to be part of a conforming culture where emphasis was on manners, cleanliness, and obedience. The mothers were generally disadvantaged in employment, housing, and health; had lower social class ratings; and had economic and emotional burdens. These findings may be due at least in part to adverse environmental circumstances and the cultural patterns and attitudes of the West Indian community. (7 refs.) - *J. K. Wyatt.*

St Mary's Hospital Medical School
London W. 2, England

2902 **OFFORD, DAVID R.; & CROSS, LUCIEN A.** Adult schizophrenia with scholastic failure or low IQ in childhood. *Archives of General Psychiatry.* 24(5): 431-436, 1971.

Adults diagnosed as schizophrenic who had low IQs in childhood had an earlier onset of psychosis and remained institutionalized longer than schizophrenics with average IQs. The 29 adult schizophrenic Ss were on the active rolls of a state hospital and had been diagnosed as schizophrenic as adults but prior to age 45. They had attended elementary or junior high school in the local, urban school system, had at least 1 IQ determination during the first 9 years of school, and had school records available through the seventh grade. They were matched for sex, age, and school class with control Ss who had attended the same school. Adult schizophrenics who had repeated a grade in school completed significantly fewer years of school than nonrepeaters, controls, or siblings. Patients with lower IQs (below 80) were hospitalized significantly earlier and accumulated significantly more institutional time than those with higher IQs. (38 refs.) - *J. K. Wyatt.*

Pennsylvania State University
Hershey, Pennsylvania 17033

2903 **VERMA, SITAPATI K.** Poverty and mental retardation. *Journal of Rehabilitation in Asia,* 12(1): 42-44, 1971.

Poverty, which is characterized by insufficient income to meet current needs, greatly limits potential physical and psychological growth and development. MR—a condition of sub-average intellectual functioning originating during the developmental period and associated with impaired adaptive behavior—is caused by many factors, but research has shown that impoverished families are more likely to have children with organically caused MR than are families at higher socioeconomic levels. The MR in these cases often results from ignorance of hazards to health and from the poor health and nutritional status of both mother and child. The child in an environment without language and cultural stimulation will exhibit retarded intellectual development, with studies showing a consistent rise in IQ accompanying higher socioeconomic status. Nutritional programs and adequate medical care for pregnant women and infants are needed to overcome this situation. (4 refs.) - *B. Berman.*

No address

2904 Institute for Research into Mental Retardation. *Infantile Autism: Concepts, Characteristics and Treatment.* Rutter, Michael L., ed. Edinburgh and London, Churchill Livingstone, 1971, 328 p., £3.50.

The publication of the proceedings of the Study Group on Infantile Autism provides a survey of recent research into autism and of current developments in concepts of this uncommon disorder. The classification of infantile autism is discussed, and findings are reported on perception and social responses, varied approaches to language, behavioral approaches to treatment, and education of and treatment for autistic children. *B. J. Grylack.*

CONTENTS: Introduction (Eisenberg); Psychoses in Childhood-A Comparative Study (Kolvin); A

Pathogenic Approach to Infantile Autism (Koupernik); A Paediatrician's Approach to Autism (Rendle-Short); Childhood Autism: A Disorder of Sensorimotor Integration (Ornitz); Visual Perception in Autistic Childhood (O'Connor); Perceptual Limitations in Autistic Children and Their Relation to Social and Intellectual Deficits (Demyer); Rules and Language (Hermelin); Spontaneous Patterns Produced by Autistic, Normal and Subnormal Children (Frith); Autism-A Central Disorder of Cognition and Language? (Rutter); Perceptual and Language Development in Autistic Children: A Comparative Study (Wing); Developmental Therapy by Parents with Their Own Autistic Child (Schopler); Theoretical and Experimental Aspects of the Behaviour Modification Approach to Autistic Children (Evans); Educational Treatment of Autistic Children (Bartak); Sensory Disorder in the Autistic Child and its Implications for Treatment (Martin); An Approach to Treatment in the Young Autistic Child (Savage); Chairman's Closing Remarks (Eisenberg).

2905 **EISENBERG, LEON.** Introduction. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 2-5.

Infantile autism raises issues of diagnosis, classification, cause, and treatment whose clarification and resolution will provide insight into many of the other unsolved clinical problems in child psychiatry. From nonrecognition, the syndrome of infantile autism has passed through overrecognition to a confusion of the disorder in the young child with schizophrenia in the adult. Knowledge of the cause of infantile autism has not advanced significantly as a result of contradictory evidence and undocumented hypotheses, and treatment of infantile autism remains elusive. (14 refs.) - *B. J. Grylack.*

2906 **KOLVIN, ISRAEL.** Psychoses in childhood—a comparative study. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 7-26.

A comparative study of family and social background, parental personalities and attitudes, evidence of cerebral dysfunction, and phenomenological and cognitive factors in infantile psychoses and late onset psychoses was undertaken to seek a preliminary clinical classification of childhood psychosis and to evolve ascertainment criteria for both psychotic groups. The main hypothesis to be tested was that psychotic disorders in childhood are dependent upon the age at which the process begins and the disorders with different ages of onset differ fundamentally. It was also hypothesized that infantile psychoses and late onset psychoses differ in etiology and phenomenology, that infantile psychotic children show different symptoms when reaching the age at which late onset psychosis begins, and that similar etiological patterns should be operative if late onset psychosis is related to adult schizophrenia. It was possible to identify features other than the ascertainment criteria differentiating the groups. These include family, genetic, and social factors, maternal personality differences, evidence of cerebral dysfunction, and phenomenological and cognitive differences. Both the main and subsidiary hypotheses were substantiated. (41 refs.) - *B. J. Grylack.*

2907 **KOUPERNIK, CYRILLE.** A pathogenic approach to infantile autism. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 27-42.

The hypothesis that infantile autism may be a more pronounced form of a disorder which is almost impossible to diagnose during infancy and which only really materializes during adolescence has not yet been proven, and EEG and related criteria have been shown to be indecisive measures for distinguishing the disorder from brain damage. Physiological research carried out in the areas of biochemistry and arousal state of the reticular system of the brainstem has not yielded any conclusive evidence. The primary conclusion that can be drawn from the anthropological-relational, organic, physiological, and cognitive approaches to autism is that they orient investigators towards comparisons by analogy with the models of congenital developmental aphasia and developmental disharmonies. (36 refs.) - *B. J. Grylack.*

2908 **RENDLE-SHORT, JOHN.** A paediatrician's approach to autism. In: Rutter,

Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 43-45.

On the basis of a study of behavioral abnormalities particularly associated with autism, 14 major manifestations of the condition have been described by pediatricians. These are difficulty mixing with other children, acting as if deaf, resistance to routine change, indication of needs by gestures, inappropriate laughing and giggling, lack of cuddling, marked physical overactivity, no eye contact, inappropriate attachment to objects, spinning of objects, sustained odd play, and standoffish manner. The presence of 7 or more manifestations indicates autism. A provisional classification of autism distinguishes two subdivisions of Group One autism: one with symptoms commencing from or soon after birth, and one in which symptoms follow some precipitating factor which occurs later in the first year of life or during the next 18 months. The Group Two autistic syndrome may develop secondarily following an organic brain lesion, deafness, blindness, or severe MR, and the Group Three syndrome may also be seen secondary to environmental stress. Since the pediatrician, unlike the child psychiatrist, sees very young infants and tends to be doctor of first referral, it is vital that he be able to distinguish these cases from other disorders and to initiate early, active treatment. (1 ref.) - *B. J. Grylack.*

2909 **ORNITZ, EDWARD M.** Childhood autism: A disorder of sensorimotor integration. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971. p. 50-68.

Childhood autism has been described as an illness of interpersonal and object relationships and as a disturbance of the comprehension and use of language. Clinical observations suggesting significant disturbances of the rate and sequence of psychomotor development and motility have indicated a possible breakdown of sensorimotor integration, and neurophysiological studies have raised the possibility of failure of the normal homeostatic balance between excitatory and inhibitory influences within the central nervous system. Research on rapid eye movement (REM) sleep has provided some evidence of a maturational delay in the differentiation of sleep EEG patterns and the development of phasic excitatory and inhibitory mechanisms during REM sleep in autistic children. The possible formulation of childhood autism as a disorder of sensorimotor integration can potentially account for not only symptoms of disordered motor expression and sensory receptivity but also those related to distortions of personality development. (50 refs.) - *B. J. Grylack.*

2910 **O'CONNOR, NEIL.** Visual perception in autistic children. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 69-79.

Studies of the free behavior of autistic children when confronted with stimuli in unrestricted fields and of free movement of their approach and avoidance to chosen displays have confirmed that autistic children show much more unmotivated nonorienting behavior than normal children and that their orienting behavior is similar to that of severely subnormal children of an equivalent MA. A series of experiments concerned with visual discrimination, sensory dominance, and motor response seemed to indicate that the autistic group failed to assign meaning to visual signals and, perhaps for this reason, failed to discriminate on the basis of visual signs. When autistic children were compared with a group of severely subnormal and a group of normal controls in eye movement studies, it was found that total visual inspection time of a presented stimulus was less in autistic children than in controls. It appears that displays are less meaningful for autistic children. The fact that these Ss make fewer comparisons of the features presented in displays is probably indicated by the smaller number of eye movements recorded for them. Relative lack of persistent cortical responsiveness in autistic children may match their same relative lack of visuomotor responsiveness as judged by eye movements. (22 refs.) - *B. J. Grylack.*

2911 **DeMYER, MARIAN K.** Perceptual limitations in autistic children and their relation to social and intellectual deficits. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 81-96.

On the basis of clinical studies, the staff of the Clinical Research Center for Early Childhood Schizophrenia has developed the new, tentative neurophysiological hypothesis that autistic and schizophrenic children who manifest their symptoms before their third birthdays have deficiencies in transferral of perceptual information from 1 sensory modality to another and to the motor system. The finding that autistic children most resemble brain-damaged children led to a comparison of the parents of brain-damaged and non-brain-damaged children, which showed that both sets of parents were alike in response to the Minnesota Multiphasic Personality Inventory (MMPI) and were essentially the same in child rearing practices, personalities, and marital interaction. While autistic children and nonpsychotic, brain-damaged children were found to be very similar in incidence of positive neurological findings, abnormal EEGs, and cross-modal transfer defects, they manifested different relationships with and responses to people. It appears that even though autistic and brain-damaged children have varieties of cross-modal and sensorimotor processing problems, the autistic and schizophrenic children have either a common visual memory deficit or a body part agnosia or both, while nonpsychotic children have either relatively better visual memories or no body part agnosia. (34 refs.) - *B. J. Grylack.*

2912 **HERMELIN, BEATE.** Rules and language. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 98-111.

Autistic children were investigated for the precise nature of their language deficits. In a study of the role of phonological features for verbal recall in 16 autistic children (mean CA 11 yrs 6 mos) and 16 normal children (mean CA 4 yrs 3 mos), it was shown that the effects of phonological structure were equal for both groups. When 12-year-old autistic and subnormal children were compared for the effects of words and sounds, autistic Ss were found to be more influenced by the position and intensity of stimuli than by their qualitative differences. Highly significant differences between normal and subnormal children as opposed to autistic Ss were also apparent in the ability to construct a series of nonverbal tasks. Despite possible indications that autistic children cannot distinguish structure or meaning from randomness, the sequences they produce follow strict and simple rules such as repetition or alternation. Though auditory-verbal memory capacity is not impaired in autistic children, the recall process depends upon an echobox type memory store, and their response patterns are relatively unrelated to and independent of the true structure of the material which they are asked to recall or reproduce. (No refs.) - *B. J. Grylack.*

2913 **FRITH, UTA.** Spontaneous patterns produced by autistic, normal and subnormal children. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 113-131.

Experiments on spontaneous production of temporal sequences were conducted with autistic children and normal and subnormal controls to determine whether the tendency among autistic children to impose on their responses patterns which had nothing to do with the actual patterns presented was independent of the input and dependent on purely internal factors. The 3 experiments consisted of card guessing, construction of a row of colored counters, and production of strings of colors and tunes on a xylophone. The patterns of the autistic group were distinguished from the patterns of the other groups in terms of strict rule adherence, restricted use of the range of elements available, and lack of originality. There was no difference in terms of pattern complexity. It appears that the tendency among autistic children for pattern composition is to some extent independent of the environment and is an expression of strong internal factors with a characteristic rigidity and stereotypy of rule application. Autistic children probably suffer from a failure in the central processing of incoming and outgoing material. This deficit involves an imbalance of internal over external factors which may account both for difficulties in language acquisition and for obsessional-ritualistic phenomena in spontaneous behavior. (11 refs.) - *B. J. Grylack.*

2914 **RUTTER, MICHAEL; BARTAK, LAWRENCE; & NEWMAN, STEVEN.** Autism—a central disorder of cognition and language? In: Rutter, Michael L., ed.

Infantile Autism: Concepts, Characteristics and Treatment. Edinburgh and London, Churchill Livingstone, 1971, p. 148-171.

Although the evidence is incomplete, it appears unlikely that infantile autism is a type of schizophrenia or is a result primarily of psychogenic influences, faulty conditioning, high physiological arousal, or social withdrawal. Recent studies have indicated, however, that autism may be due to the interaction between familial autistic traits and brain damage, may be a response to language and perceptual abnormalities, and that intellectual retardation is an important aspect of the disorder. Clinical, comparative, and experimental investigations of autistic children of all levels of intelligence have demonstrated the presence of a severe and extensive defect in language comprehension and in central functions associated with language and with the processing of symbolic or sequenced information. While it is uncertain whether the language/cognition defect is sufficient in itself to lead to autism and whether some interaction with particular personality attributes or familial environment is necessary, the consistent finding of gross language and cognitive defects and the failure to identify deviant factors in the environmental situation indicate that a cognitive handicap involving language is the main factor involved in the pathogenesis of autism. (70 refs.) - *B. J. Grylack.*

2915 **WING, LORNA.** Perceptual and language development in autistic children: A comparative study. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 173-197.

The perceptual disorders, language abnormalities, and secondary behavioral problems of autistic children from infancy until ages 7 to 10 were compared with those of other handicapped and normal children. Ss were 20 children with early childhood autism with some useful speech, 7 children with early childhood autism with no speech, 11 children with receptive aphasia with some useful speech, 10 children with executive aphasia with some useful speech, 15 partially blind/partially deaf children, 15 children with Down's syndrome with some useful speech, and 25 normal children. The language problems of autistic Ss were very similar to those of the aphasic Ss. Visual perceptual problems and difficulties with nonverbal language among autistic Ss closely resembled these disorders in the partially blind/partially deaf Ss. The most striking abnormality in most of the autistic Ss and all of the partially blind/partially deaf children was a lack of interest in the surrounding environment. All handicapped groups manifested early signs of abnormal development in babyhood, and all showed improvement in 1 or more behavioral categories with increasing age. A theory of multiple cognitive and language dysfunctions may account for similarities and differences between the groups of children. (19 refs.) - *B. J. Grylack.*

2916 **SCHOPLER, ERIC; & REICHLER, R. J.** Developmental therapy by parents with their own autistic child. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 206-227.

Many of the parents who participated in developmental therapy with their own autistic children in the 5-year Child Research Project have developed a high degree of objectivity, investment, and skill and have extended their capacity to bring more meaningful organization into the lives of their children. In this approach, parents were utilized as the primary developmental agents to prevent the elaboration of psychotic behavior with its debilitating effects on the child and his family, to achieve maximum adaptation between the child and his family or community, and to promote recovery where possible. Although the project is only in its second year, several trends for change in the children can be identified. For some, a recovery from autism to relatively normal functioning can be predicted. For others, the psychotic behavior, screaming, self-destructive behavior, and lack of relatedness are no longer a problem. In all families, realistic appraisal of the child by parents contributed to making their best energies available to him while either maintaining or improving the adjustment in equilibrium between all family members. (37 refs.) - *B. J. Grylack.*

2917 **EVANS, IAN M.** Theoretical and experimental aspects of the behavior modification approach to autistic children. In:

Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 229-251.

As the autistic syndrome becomes better understood, functional analysis can become more profound, and behavior will probably be shown to be a function not only of specific stimuli but also of more complex conditions, including defects within the autistic child. In evaluating treatment, it is important to distinguish between intrinsic and extrinsic aspects. The 2 most common experimental designs are the reversal and multiple baseline techniques. A related problem in specific behavior manipulation is the immediate benefit to the child. The one clear treatment goal that can be identified objectively is the ability of the child to lead a life within society. Thus, the institution is not a bad standard of improvement. (61 refs.) - *B. J. Grylack.*

2918 **BARTAK, LAWRENCE; & RUTTER, MICHAEL.** Educational treatment of autistic children. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 258-280.

A preliminary follow-up examination of 50 autistic children was undertaken to assess the utility of 3 approaches to special educational treatment of these children. Eight children in Unit A were instructed with regressive techniques, with minimal attention directed towards the teaching of specific skills. Eighteen children in Unit B received treatment in a more permissive classroom environment which combined special education methods with regressive techniques. Twenty-four children in Unit C attended a school for autistic children only, which exemplified the approach in which perceptual, motor, and cognitive handicaps are viewed as primary. Various indications of approval by the staff in all 3 units almost always followed acceptable behavior by the children. Deviant behavior elicited staff disapproval expressed with a critical tone in Unit C, whereas it was equally likely to be followed by gentle approval or disapproval in Unit A. Unit B was intermediate. Although the follow-up period has been too short to provide meaningful comparisons between the 3 groups, the educational progress of the total group has been encouraging: there was considerable improvement in responsiveness, a third of the mute children began to speak, the number who could read doubled, and 10 percent were transferred to normal schools. Reading comprehension lagged significantly far behind accuracy, however. (39 refs.) - *B. J. Grylack.*

2919 **MARTIN, J. A. M.** Sensory disorder in the autistic child and its implications for treatment. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 286-296.

Autism appears to be at least 2 separate and distinct syndromes, as indicated by analysis of the hearing behavior of autistic children. A feature common to certain groups of children who are late in learning to talk, excluding those with peripheral hearing loss, is the considerable difficulty experienced in arriving at an accurate evaluation of their hearing. In the instance in which there is a decision not to respond, there is an activity of will resulting in the withholding of response (impaired social response). In other situations, the judicative ability of the autistic child may be impaired (indecisive response). The failure of the autistic child to distinguish complex sounds indicates failure of sensory organization, which may result in imperfect multisensory integration. Insufficient interaction with the mother due to inadequate care on her part or to a neurological deficit in the child himself, complicated by some degree of MR, is a significant form of deprivation of the autistic child. Failure in the acquisition of spoken language, and perhaps aspects of MR itself, might be considered an inevitable consequence of severe sensory disorganization. (24 refs.) - *B. J. Grylack.*

2920 **SAVAGE, VALERIE A.** An approach to treatment in the young autistic child. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 297-307.

Although autistic children may have speech difficulties, it seems probable that their inability to handle symbolic form and to classify a set of

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symbols into the language system of their culture may be the primary factor in their total cognitive disability. Rather than overwhelming the child with stimuli, through the various sense modalities, which he cannot organize for himself, it appears more logical to adopt a unisensorial approach. The aim of such an approach is to select a single modality, help the child to comprehend stimuli received through this modality, and then introduce a second modality into the teaching situation with a view towards integrating the impressions into a meaningful whole. Any therapeutic program in the preschool years must consider the particular difficulties encountered by parents of the severely handicapped child as well as the ultimate educational placement of the child and must form a base from which the child can progress to more formal learning. (21 refs.) - *B. J. Grylack.*

2921 **EISENBERG, LEON.** Chairman's closing remarks. In: Rutter, Michael L., ed. *Infantile Autism: Concepts, Characteristics and Treatment.* Edinburgh and London, Churchill Livingstone, 1971, p. 313-315.

While fundamental knowledge of infant development and childhood autism remains extremely limited and hope for prevention of autism may be premature, clarification of terminology may promote better understanding of the cultural implications of the disorder. The term 'autism' should be applied only to disorders with an onset not later than 30 months. The term itself is inaccurate, for it implies a retreat from a prior orientation to reality, whereas autistic children cannot relate and never have. Although there is no behavior that cannot be changed, the extent of progress varies greatly from individual to individual. Parents of autistic children will be spared great emotional cost if they are protected from false promises and are encouraged to expect only modest accomplishments from the treatment of these children. (No refs.) - *B. J. Grylack.*

2922 **BROWN, ROY E.; & HALPERN, FLORENCE.** The variable pattern of mental development of rural black children. *Clinical Pediatrics,* 10(7): 404-409, 1971.

The effects of deprived environment and malnutrition on the mental development of rural Black children in the first 3 years of life were studied. Physical, medical, and psychologic (Gesell Developmental Scales) examinations were given to 402 Black children aged 1 week to 36 months. In height and weight the Ss were below the reference standards, and almost all were anemic. The Gesell Scales, which examine mental functioning in 4 areas (motor, adaptive, personal-social, and verbal), yield a Developmental Quotient (D.Q.) score, with 100 as the standard. D.Q.s of Ss were 117 at 1 week to 3 months, 101 at 3-6 months, 96 at 16 months, 90 at 19 months, and 86 at 36 months. Verbal ability fell off early and showed the most retardation by 36 months. Significant relationships were found between child's D.Q. and parents' educational level, and between D.Q. and age. No significant correlation was found between D.Q. and suboptimal growth, or between anemia and D.Q., suggesting that Ss' limitations were not due entirely to malnutrition, but at least in part to impoverished environment. Ss' D.Q.s up to the age of 15 months suggest that Black intellectual potential is as good as or better than that of the White population. (12 refs.) - *K. L. Woodworth.*

Mount Sinai School of Medicine
New York, New York 10029

2923 **HERMELIN, B.; & O'CONNOR, N.** Spatial coding in normal, autistic and blind children. *Perceptual and Motor Skills,* 33(1): 127-132, 1971.

Ten autistic, 10 blind, and 20 sighted children, 10 of whom were blindfolded for the experiment, as well as 10 blindfolded adults, were tested to determine whether tactile stimulation would be coded in terms of visual location, even when no visual cues were available. Stimulation was administered in a predetermined random order, with the provision that each tactile stimulus-verbal response pair occurred equally over a series of 40 trials. CA, sensory impairment, and psychiatric diagnosis did not seem to be particularly relevant in the determination of response strategy. Ss who could see what was done gave predominantly location responses after hand reversal, regardless of whether they were normal or autistic. In contrast, Ss who carried out the task without vision responded mainly in terms of fingers touched, regardless of whether they were blind or blindfolded and adults or children. It appears that most Ss responded to

immediate sensory input rather than to internally represented schemata of visually organized space. (11 refs.) - *B. J. Grylack.*

Drayton House, Gordon Street
London W. C. 1, England

2924 **VALENTE, MARIO.** Autism: Symptomatic and idiopathic—and mental retardation. *Pediatrics,* 48(3): 495-496, 1971. (Letter)

Autism is a constellation of behavioral symptomatology including self-stimulatory activity as well as deficiencies in socialization skills, play activity, verbal communication, and self-help skills. In most cases, no cause can be found either on the basis of historical data or on the basis of laboratory tests and careful neurological examination. Autism should not be considered an emotional disorder amenable to therapy in contrast to considering MR as an organic disorder not amenable to therapy. Autism should be interpreted in the majority of cases as a form of MR with a severe prognosis. (No refs.) - *B. J. Grylack.*

UCLA Center for the Health Sciences
Los Angeles, California 90024

2925 **O'GORMAN, GERALD.** *The Nature of Childhood Autism.* Second edition. New York, Appleton-Century-Crofts, 1970, 163 p. $7.75.

In the present volume, autism is treated as a symptom seen in a number of conditions and is operationally described as a selective or total withdrawal from the environment without true sensory deficits. Ten photographs of autistic children alone and in group play as well as a discussion of views about etiology, pseudoschizophrenic syndromes, and treatment are included. Seventeen case examples of hospitalized children are interspersed throughout the text. (60 refs.) - *B. J. Grylack.*

2926 **BENDER, LAURETTA.** Autism in children with mental deficiency. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 49-55.

Autism is not synonymous with psychosis nor does it indicate a specific type of mental illness. A review of the history of the concept of autism reveals the value of Kanner's syndrome of early infantile autism. But this designation is limited to a particular group of children and does not constitute a clinical or etiological entity. Autistic thinking and actions are a primitive form of behavior which may either persist or be reverted to as a defense mechanism against other pathologies. The term only describes a form of behavior and does not indicate either the cause or ultimate outcome. (19 refs.) - *S. Glinsky.*

Creedmoor State Hospital
Queens Village, New York

2927 **MENOLASCINO, FRANK, J.** The facade of mental retardation: Its challenge to child psychiatry. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 95-105.

The child psychiatrist is in an ideal position to employ present clinical and theoretical knowledge relative to psychosocial factors producing MR to treat and prevent apparent MR. The Ss were 616 children suspected of having MR, aged seven days to eight years, who were evaluated by a full clinical team including a psychiatrist between 1958 and 1963. One hundred and ninety-one had prominent psychiatric problems, of whom 40 had disturbances without MR while 151 had both emotional disturbances and MR. Four mechanisms which gave a clinical facade of MR in the group of 40 were inhibition of learning by chronic anxiety, suppression of curiosity, inhibition of intellectual functions by conflict regression processes, and the presence of regressive emotional disturbances. These findings suggest that the bulk of children presently designated as mildly retarded is not fixed at this level of intellectual functioning. This facade of MR is one of the most promising areas of investigation and progress for the child psychiatrist. (41 refs.) - *S. Glinsky.*

University of Nebraska College of Medicine
Omaha, Nebraska

2928 **REISER, DAVID E.** Psychosis of infancy and early childhood as manifested by children with atypical development. In: Menolascino, Frank, J. ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 57-82.

Psychosis of infancy, which is generally not well understood among physicians, is a grave disturbance in the preschool years which can be diagnosed from characteristic history and clinical signs in the child as well as from certain parental history. Knowledge of the developmental history of the child is important in differential diagnosis. Diagnostic categories to be considered are: retardation, chronic brain syndromes of various origin, psychologic regressive reactions, infantilization, deafness, aphasia and childhood schizophrenia. The fragmented personality permits single well developed functions, but not integrated ensembles. Psychosis of infancy represents regression to a level of omnipotence and isolation which actively excludes the outside world. Aspects of parent-child relationships that could make the child vulnerable to such development are discussed. To reverse the isolation, treatment through intensive psychotherapy, reeducation and parental participation has been developed. (149 refs.) - *S. Glinsky.*

James Jackson Putnam Children's Center
Roxbury, Massachusetts

2929 **CALL, JUSTIN D.** Prevention of autism in a young infant in a well-child conference. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 322-328.

If early autistic trends are diagnosed and treated early enough, reversal can occur. A case of a male infant, seen at a well-baby clinic between the ages of 2½ months and 27 months, illustrates the pertinence of mother-infant interaction in the production of autistic trends in infancy. The child presented with autistic tendencies and symptoms, which were connected with efforts by the mother to distance herself from the child. This affective defect or interlocking affective freeze was gradually dissipated through sessions with the mother, who was encouraged to exhibit affection toward the child and to express aggressive tendencies caused by experiences with an older child. The mother's ability to revitalize her mothering role with supportive educational help from the physician was the prime factor producing change. (5 refs.) - *S. Glinsky.*

University of California School of Medicine
Los Angeles, California

MEDICAL ASPECTS – Convulsive Disorders

2930 **PRENSKY, ARTHUR L.; DEVIVO, DARRYL C.; & PALKES, HELEN.** Severe bradykinesia as a manifestation of toxicity to antiepileptic medications. *Journal of Pediatrics,* 78(4): 700-704, 1971.

A case is reported in which an epileptic patient suffered a progressive deterioration in performance and behavior over a 22-month period, associated with a medication-induced bradykinetic state complicated only briefly by an episode of lethargy and ataxia. The S, a 9-year-old male, was first seen in 1964 after a series of "trances" over a 1-year period. These increased in frequency to 20-30 per day. Treatment with phenobarbital and ethosuximide caused cessation of seizures, and the S was controlled for 3½ years on medication consisting of the addition of paramethadione and substitution of diphenylhydantoin (Dilantin) for the phenobarbital. Three years later the S had a grand mal seizure, minor seizures which were difficult to control, and other speech and movement symptoms which worsened over a 20-month period. Dilantin dosage was reduced without alteration of this bradykinetic state. Dramatic improvement occurred only when the S was placed on a ketogenetic diet and removed from substantial quantities of medication. The S was hospitalized

on 2 occasions to determine the possibility of an underlying degenerative disease of the brain. The assessment of this case points to the likelihood that the progressive bradykinesia is due to the toxic effect of a combination of antiepileptic drugs rather than the result of any excessive single medication. (13 ref.) - *M. S. Fish.*

St. Louis Children's Hospital
St. Louis, Missouri 63110

2931 **GREEN, JOSEPH B.; & HARTLAGE, LAWRENCE C.** Comparative performance of epileptic and nonepileptic children and adolescents (on tests of academic, communicative and social skills). *Diseases of the Nervous System*, 32(6): 418-421, 1971.

A study to measure among epileptic and nonepileptic children similarities and differences in the development of academic, social, and communicative skills has shown that epileptic children scored lower in school achievement and language usage. The study population was 50 randomly selected epileptic children and adolescents (mean age of 11.4 years) with intellectual abilities ranging from superior to mildly retarded (mean IQ of 81). Diagnosis of the Ss was: 34% with grand mal attacks; 32% with petit mal; 20% with focal motor or sensory seizures; and 14% with psychomotor seizures. Comparison with nonepileptic normative controls by use of the Wide Range Achievement Test, the Vineland Social Maturity Scale, and the Mecham Language Usage Test showed that, in comparison to the norms, epileptics were superior in terms of self direction and were equal on general self help skills. Greatest discrepancies were in occupation and locomotion, where epileptics scored significantly lower. They were consistently but not significantly lower on socialization and communication. Their use of language was quite low, but reading ability was least impaired. Arithmetic performance was below the expected level. One explanation of these differences may be the possibility that for epileptics, parental expectancies may be lower, and this may hinder the epileptic in the realization of his capabilities. Emphasis of his assets and responsibilities might result in better achievement of potential levels. (8 refs.) - *M. S. Fish.*

Indiana University Medical Center
Indianapolis, Indiana 46202

2932 **GELLER, MARTIN; & *CHRISTOFF, NICHOLAS.** Diazepam in the treatment of childhood epilepsy. *Journal of the American Medical Association*, 215(13): 2087-2090, 1971.

Diazepam (Valium) was used for long-term control of seizures (median 2 years) in 38 epileptic children. Eleven of 16 patients with petit mal had complete suppression of attacks, and 3 others had partial suppression. Fourteen of 20 children with myoclonic and akinetic seizures showed substantial improvement. Five of 12 children with psychomotor attacks had an excellent response. Improvement occurred in EEGs as well as in clinical manifestations. Since tolerance for diazepam varied greatly, dosage should be increased to a point of control or intolerance. No severe side effects were noted. Diazepam appears to be the treatment of choice for petit mal and minor seizures of childhood. (18 refs.) - *E. L. Rowan.*

*79-01 Broadway
Elmhurst, New York 11373

2933 **DAWSON, K. P.; & JAMIESON, A.** Value of blood phenytoin estimation in management of childhood epilepsy. *Archives of Disease in Childhood*, 46(247): 386-388, 1971.

An initial estimation of blood phenytoin levels in 30 epileptic children (CA range 6 to 10 yrs) who had received phenytoin for 1 month to 5 years revealed that only 25% of the children had therapeutic blood levels. Ten of the 23 children with low blood levels were free of convulsions. Patients were seen at 2-week intervals until blood levels neared the therapeutic range and then once a month. Under this close supervision therapeutic blood levels rose to 80%, and 11 of 13 children who initially had convulsions and were outside the therapeutic range were free from convulsions after 6 months. The 2 children who failed to attain therapeutic blood levels continued to have convulsions. Poor initial blood levels in this series were due to prescriptions of inadequate dosage and patient failure. (4 refs.) - *J. K. Wyatt.*

Stobhill General Hospital
Glasgow N. 1, Scotland

2934 **LIPMAN, IVAN J.; ISAACS, EDWARD R.; & SUTER, CARY G.** Petit mal status: A variety of non-convulsive status epilepticus. *Diseases of the Nervous System*, 32(5): 342-345, 1971.

The forty-first documented case of PMS (petit mal status epilepticus)—a 40-year-old retarded woman—demonstrates that PMS may often go undetected and requires more awareness by physicians of its frequent occurrence in adults. Clinical diagnosis can only be suspected; accurate diagnosis depends on EEG analysis (continuous or sub-continuous generalized spike-wave complexes is diagnostic). Mental state in PMS is altered and S appears groggy, sedated, drowsy, or lethargic. There are apparently precipitating factors, most notably menstruation. Victims are not a homogeneous group, but part of a large class with prolonged, non-conclusive epileptic attacks. Intravenous diazepam and trimethadione are favored control agents. (22 refs.) - *B. Berman.*

Medical College of Virginia
Richmond, Virginia

2935 **VULLIAMY, DAVID.** Unwanted effects of anticonvulsant drugs. *Developmental Medicine and Child Neurology*, 13(1): 107-109, 1971.

The treatment of epilepsy with anticonvulsant drugs is often accompanied by toxic effects; however, since several of these appear to be long-term in nature, differentiation between the effects of drug and deterioration due to the fits themselves is often difficult. Investigations in this area have not resulted in agreement regarding the effects of the drugs as opposed to effects of the illness. One investigator claims that while drugs like phenobarbitone may affect a child psychologically, most of the permanent slowness is the result of the disorder which caused the epilepsy. Other investigators claim that the three most commonly used epileptic drugs produce metabolic changes which, in turn, cause more widespread CNS dysfunction. The use of phenytoin has been associated with side effects such as drowsiness, behavior disturbance, ataxia, nystagmus, dizziness, blurred vision, diplopia, incontinence of urine, and exacerbation of fits. Disturbance of folic acid metabolism has been detected in most epileptics treated with drugs over prolonged periods of time. The mechanism and clinical consequences of this effect, however, are unknown. Ethosuccimide has been associated with significant lowering of IQ, and correlations between the use of anticonvulsant drugs and impairment of mental function have been suggested. Other drugs can affect blood levels of certain anticonvulsant drugs. Vitamin D utilization and serum calcium levels have been shown to be affected by antiepileptic materials. As a result of these and other recently reported data, the clinician must exercise considerable care in the use of these drugs. (13 refs.) - *M. S. Fish.*

Dorset County Hospital
Dorchester, Dorset, England

2936 **KILLIAN, JAMES M.; & FROMM, GERHARD H.** A double-blind comparison of nitrazepam versus diazepam in myoclonic seizure disorders. *Developmental Medicine and Child Neurology*, 13(1): 32-39, 1971.

Nitrazepam and diazepam in combination with phenobarbital are of equal effectiveness at maximum tolerated doses in the treatment of patients with drug-resistant myoclonic seizure disorders. Of 9 patients (5 male and 4 female), ranging in age from 2½ to 17 years (7 under 10 years of age) with poorly controlled or uncontrolled myoclonic, minor motor, or akinetic seizures, 8 were MR (3 SMR). Seizures were moderate (2-10/day) in 7 and severe (above 10/day) in 2. Diazepam (10-20 mg/day) and nitrazepam (5-10 mg/day) were given the Ss initially in a sequential double-blind manner, and all Ss received 30-60 mg/day of phenobarbital, but no other medication. Medication was gradually increased to either maximum clinical control or to where toxic side effects occurred. At 6 months the Ss were crossed over to the second medication and continued for 6 more months. Evaluation of the Ss (laboratory tests, EEG's, clinical response) showed that both drugs afforded more than a 50% improvement in seizure control in 5 cases. Complete control resulted initially for 4 Ss (2 on each drug); however, none of the cases remained completely free of seizures for the entire year. In 4 Ss no essential change was noted at 1 year compared to pretreatment status. None of the 8 MR Ss showed improvement in mental status beyond that normally expected with increase in age. EEG counts correlated well with clinical seizure history. The principal side effects were

transient drowsiness and hypotonia. High doses of diazepam were associated with respiratory symptoms for 3 Ss. Although both drugs were equal in effectiveness, approximately twice the total dosage of diazepam (compared to nitrazepam) was required. (12 refs.) - *M. S. Fish.*

Baylor College of Medicine
Houston, Texas

2937 **TAYLOR, D. C.; FALCONER, M. A.; BRUTON, C. J.; & CORSELLIS, J. A. N.** Focal dysplasia of the cerebral cortex in epilepsy. *Journal of Neurology, Neurosurgery and Psychiatry*, 34(4): 369-387, 1971.

On the basis of microscopic examination of lobectomy specimens from 8 male and 2 female chronic epilepsy patients, a particular structural abnormality was defined and evaluated in terms of its bearing on clinical findings. Six of the patients underwent a temporal lobectomy, 3 had an anterior frontal ablation, 1 an occipital lobectomy, and 1 a parietal gyrectomy (1 patient had 2 operations). In all cases, the cortical surface of the fixed lobe or tissue appeared normal to the naked eye, and in 7 of the 10 specimens there was nothing remarkable about the cut surface. The most striking microscopic feature was the localized disruption of the normal cortical lamination by an excess of large aberrant neurons scattered randomly through all but the first layer. The giant neurons were approximately the size of a giant Betz cell. The abnormalities were interpreted as a particular form of localized cortical dysplasia in which anomalous populations of neurons and often glia underlie the electrical and clinical manifestations of certain focal forms of epilepsy. (19 refs.) - *B. J. Grylack.*

Park Hospital for Children
Headington, Oxford, England

2938 **NORRIS, J. W.; & PRATT, R. F. A.** A controlled study of folic acid in epilepsy. *Neurology*, 21(6): 659-664, 1971.

A controlled drug trial was performed on 51 epileptic outpatients by comparing the number of fits occurring during a 3-month period of folic acid administration to those occurring during placebo administration. Results showed no significant change in seizure frequency in either the folate-deficient or borderline folate-deficient group or in both combined following folic acid administration ($p<0.10$). During placebo or folate administration, patients often volunteered statements concerning improvement or deterioration of seizure frequency, but examination of the card on which the number of seizures was listed rarely confirmed that impression. It appears that folate deficiency in epileptics should be treated as it is in nonepileptics. Furthermore, since the effects of folate deficiency can be serious, there should be no hesitation in prompt treatment of folate deficiency in epileptics. Longer duration of treatment and greater numbers of patients may be necessary to prove any definite effect of folic acid administration on epilepsy. (35 refs.) - *B. J. Grylack.*

University of Toronto Clinic
Toronto, Ontario, Canada

2939 **BOOKER, H. E.; TORMEY, A.; & TOUSSAINT, J.** Concurrent administration of phenobarbital and diphenylhydantoin: Lack of interference effect. *Neurology*, 21(4): 383-385, 1971.

Serum drug levels were determined in 20 Ss in an epilepsy clinic who were taking diphenylhydantoin (DPH) and 66 Ss who were taking both DPH and phenobarbital in an attempt to measure the possible effect of phenobarbital on serum DPH levels and the correlation between the levels of the 2 drugs. In addition, 8 institutionalized patients who had been receiving DPH at a stable dose chronically had barbiturates introduced experimentally into their regimen. In the outpatients, while the mean DPH level was higher for both the high- and low-phenobarbital groups, none of the differences was statistically significant. A small increase in DPH level was seen in the 8 inpatients during the 2-week phenobarbital period, but it was not significant. These data confirm other observations that the interference of phenobarbital with DPH metabolism as noted in animals or in single-dose studies in man is of little, if any, clinical significance when the drugs are taken on a chronic basis, as is the case with epilepsy. (10 refs.) - *B. J. Grylack.*

Neurological and Rehabilitation Hospital
Madison, Wisconsin 53704

2940 **FERNANDEZ, HERIBERTO; McKINNEY, WILLIAM; & SPUDIS, EDWARD V.** Pulsatile echoencephalography: Correlations with K-complexes and epileptic discharges. *Neurology*, 21(3): 225-231, 1971.

Preliminary results of simultaneous EEG, echo, and blood pressure recordings were obtained in approximately 30 normal Ss and 25 epileptics in an attempt to establish correlations with K-complexes and epileptic discharges by means of pulsatile echoencephalography. The recordings were made during several varieties of epileptic attacks and also during periods of auditory stimulation which evoked K-complexes; these K-complexes were related in a definite way to changes in blood pressure. Most epileptic discharges could not be evaluated by echoencephalography, because they were associated with disruptive movements of the head of the patient. However, alterations in some pulsatile echoes were noted in some of the epileptic patients as a result of seizures. Further studies must be carried out with a larger series of patients with frequent seizures in order to clarify the topography of the echo change. (5 refs.) - *B. J. Grylack.*

Bowman Gray School of Medicine
Winston-Salem, North Carolina 27103

2941 **BOWE, J. C.; *CORNISH, E. JEAN; & DAWSON, MOLLIE.** Evaluation of folic acid supplements in children taking phenytoin. *Developmental Medicine and Child Neurology*, 13(3): 343-354, 1971.

The administration of phenytoin as an anticonvulsant also results in low concentration of folate in the blood. Twenty-eight epileptic children received folic acid supplementation in a double-blind crossover study in order to determine changes in the incidence of fits, EEG patterns, behavior, and gum hyperplasia. Despite persistently high folate levels after 3 months of therapy there was no significant change in the parameters under study. The anti-folate and anticonvulsant effects of phenytoin do not appear to be related. (28 refs.) - *E. L. Rowan.*

*Victorian College of Pharmacy
Parkville, Victoria 3052, Australia

2942 **STORES, GREGORY.** Cognitive function in children with epilepsy. *Developmental Medicine and Child Neurology*, 13(3): 390-392, 1971.

Children with epilepsy need to be evaluated not in terms of multifactorial global intelligence but in specific cognitive functions. Temporal lobe lesions in the dominant hemisphere are associated with impaired verbal ability, temporal lobe lesions in the nondominant hemisphere with performance and memory defects, and centrencephalic lesions with poor attention span. Localization of such specific defects can result in better re-educational methods and better forms of cognitive assessment are needed. (30 refs.) - *E. L. Rowan.*

St. James' Hospital
Leeds, England

2943 **BUCHANAN, ROBERT A.; & ALLEN, RICHARD J.** Diphenylhydantoin (Dilantin) and phenobarbital blood levels in epileptic children. *Neurology*, 21(8): 866-871, 1971.

A clinical study of diphenylhydantoin (Dilantin) blood concentrations was performed on 203 epileptic children in an attempt to relate Dilantin plasma levels to dose, seizure control, and side effects in children. One hundred and twenty-eight Ss were taking phenobarbital, and many were receiving other medication. Dilantin and phenobarbital levels were measured by a gas chromatographic procedure. In the group receiving Dilantin without phenobarbital, 54% achieved 100% control, and 22% had less than 50% control or no control. In the group receiving both medications, 54% experienced 100% control, and 21% had less than 50% control or no control. In the Dilantin group, both the daily dosage and blood concentration appeared to be slightly greater in the patients with incomplete control. In the Dilantin and phenobarbital group, the phenobarbital concentration seemed fairly uniform. Gingival hyperplasia, the main side effect of Dilantin, did not appear to be dose-related. Phenobarbital was not found to exert any effect on Dilantin blood levels in this study. (16 refs.) - *B. J. Grylack.*

Parke, Davis & Company
Ann Arbor, Michigan 48106

2944 **HIRSCH, CHARLES S; & MARTIN, DENNIS L.** Unexpected death in young epileptics. *Neurology*, 21(7): 682-690, 1971.

Of 58 epileptics (ages 6 to 30 years) who died unexpectedly, 19, for whom comprehensive records were available, showed pathophysiological mechanisms apparently related to acute disruptions of brainstem cardiac or respiratory function, or both, resulting from seizure discharge. Five of these were MR. No instance of obstructive aspiration was observed. Of 8 fatalities witnessed, 4 were incidental to grand-mal seizures. Complete gross and microscopic autopsy revealed no lethal autonomic lesion in any case. The 58 cases were about half the total number of deaths attributed to epilepsy for the age range in Cuyahoga County, Ohio, 1950-59. (20 refs.) - *B. Berman.*

Case Western Reserve University
Cleveland, Ohio

2945 **AICARDI, JEAN; & BARATON, JANINE.** A pneumoencephalographic demonstration of brain atrophy following status epilepticus. *Developmental Medicine and Child Neurology*, 13(5): 660-667, 1971.

Repeated pneumoencephalograms (PEGs) were done in the case of 15 infants and children hospitalized for status epilepticus. The first PEG was performed usually within 10 days of the onset of status and the second within 2 months in most instances. The 15 patients were subdivided into group I, comprised of the 9 patients in whom status was the first convulsive episode, and group II, consisting of the 6 children with previous chronic epilepsy. In 13 out of 15 cases (group I, 7 cases; group II, 6 cases), unilateral or bilateral ventricular dilation appeared *de novo* or as a gross increase in a previously slightly abnormal ventricular size. There was good correlation between the side of the brain showing postconvulsive atrophy in the PEGs and the side affected predominantly or exclusively by seizures, atrophy being located or predominating in the hemisphere opposite the side affected by the seizures in all but 1 case of localized status and being generalized in the patients in whom the fits had been bilateral. The ventricular dilation in cases of status epilepticus appears to be due to atrophy and not to localized or generalized hydrocephalus. (14 refs.) - *B. J. Grylack.*

Hopital St. Vincent-de-Paul
Paris XIV^{e}, France

MEDICAL ASPECTS – Chromosomal

2946 **JOHNSON, CHARLES F.** The thumb in trisomy-18. *Journal of the American Medical Association*, 215(3): 489, 1971. (Letter)

The unusual appearance of the thumb in cases of trisomy 18 can adequately distinguish this condition from others which exhibit similar hand anomalies. In trisomy 18 the thumb is high set and short with an ulnar clinodactyly which may cause overlapping of the second digit. This aspect and the marked hypoplasia of the thenar eminence are unique to trisomy 18. (2 refs.) - *N. Mize.*

No address

2947 **SPENCER, D. A.** Down's syndrome: Sex difference in relation to maternal age. *Lancet*, 1(7713): 1356, 1971. (Letter)

A new study lends support to previous accounts which have noted a significantly lower mean maternal age for female mongoloids than for male mongoloids. In a sample of 89 hospitalized inpatients with Down's syndrome (40 female, 49 male) in the Leeds (England) area, the mean maternal age for the female Ss was 31.55 years, significantly lower than the 34.55 years recorded for male Ss. (No refs.) - *N. Mize.*

Meanwood Park Hospital
Leeds, England

2948 **LARGEY, GALE P.; & LARGEY, KATHLEEN A.** Down's syndrome: Sex difference in relation to maternal age. *Lancet*, 1(7711): 1242, 1971. (Letter)

Recent studies correlating the maternal age at birth of a mongoloid child with the sex of the affected child have revealed significant differences in mean maternal age. Of 74 cases (39 female, 35 male) examined between 1963 and 1969, mean maternal age at birth of a female mongoloid child (33.28 years) was significantly lower than that for males (36.25 years). This finding is in contrast to reported results of previous studies and suggests that further examination of chromosomal aberrations associated with Down's syndrome may reveal some sex-linked factors. (4 refs.) - *N. Mize.*

Mansfield State College
Mansfield, Pennsylvania 16933

2949 **McQUARRIE, HOWARD G.; SCOTT, CHARLES D.; ELLSWORTH, HOMER S.; HARRIS, JOHN W.; & STONE, RODNEY A.** Cytogenetic studies on women using oral contraceptives and their progeny. *American Journal of Obstetrics and Gynecology*, 108(4): 659-665, 1970.

To determine the incidence and type of various chromosomal aberrations in healthy females taking oral contraceptive pills and their progeny, cytogenetic study of peripheral blood lymphocyte cultures was done on (1) 50 babies born to mothers who had previously been using sequential mestranol and mestranol with chlormadinone; (2) 23 women who had had normal births prior to this study and who were currently using oral contraceptives; and (3) a control group of 20 women who had never used any type of steroidal contraceptive. In addition dermatoglyphics were analyzed on most of the Group 1 progeny. In all cases but one (a D/G translocation mongoloid) the dermatoglyphics and the *atd* angle fell within normal range. Increased chromosome breakage and satellite association were observed in the 23 women using oral contraceptives over the 20 Ss on no gestagen therapy. Further cytogenetic studies are needed to clarify the significance of chromosome aberrations as an indicator of the sensitivity of cellular change to the contraceptive agent used. (8 refs.) - *J. C. Moody.*

Western Gynecological and Obstetrical Clinic, Inc.
Salt Lake City, Utah 84102

2950 **DUSSAULT, R.; DESTINE, M. L.; & DALLAIRE, L.** Etude cytogenetique de 440 patients institutionalises pour retard psychomoteur (Cytogenetic study of 440 patients institutionalized for psychomotor retardation). *Canadian Psychiatric Journal*, 16(3): 253-259, 1971.

Chromosomal anomalies in an MR population corresponded with observed psychomotor defects and the mean age of the MRs. Chromosome analysis was done on 440 patients in an MR institution. Among the 88 patients who had Down's syndrome, 5 had a reciprocal translocation, producing a frequency of 5.6%. The parents of 4 of these children had a normal chromosomal pattern. Of the remaining 352 patients, 2 had a gonosomal anomaly, producing a frequency of 1.4%. These data are compatible with the nature of the psychomotor defects observed and the mean age of the patients. The Ss constituted a heterogeneous group of MR individuals. (36 refs.) - *S. Glinsky.*

Universite de Sherbrooke
P. Quebec, Canada

2951 **MORRIS, ROSE.** Down's syndrome in New Zealand. *New Zealand Medical Journal*, 73(467): 195-198, 1971.

A survey of Down's syndrome in New Zealand showed variations according to maternal ages for Europeans but not for Maoris, suggesting that the older Maori mother is relatively immune to Down's syndrome. One case of a European family was found in which 2 Down's syndrome siblings were associated with 20/21 trisomy, the maternal ages being 39 and 41 years. The results suggest that Down's syndrome is a European disease, and recent findings of abnormal palmar ridge patterns among Down's syndrome families lend support to this conclusion. Eradication of the syndrome will depend on routine fetal chromosome analysis in all pregnancies. This procedure would be enhanced if simpler analysis techniques were available. (3 refs.) - *S. Glinsky.*

Department of Health
Hamilton, New Zealand

2952 **SCHMID, W.** Erbberatung bei Chromosomenanomalien (Genetic counseling in cases of chromosomal anomalies). *Monatsschrift fur Kinderheilkunde*, 119(4): 158-160, 1971.

Genetic counseling is discussed in connection with the autosomal trisomy syndromes, particularly the cytogenetic variants of Down's syndrome. The appearance of any number of symptoms justifies the search for structural chromosomal anomalies in retarded children or in children with multiple malformations. In the case of hereditary transmission of chromosomal anomalies, amniocentesis makes possible a prenatal diagnosis. (No refs.) - *S. Glinsky.*

Genetisches Forschungslabor der
Universitats-Kinderklinik
CH-8032 Zurich, Switzerland

2953 **PAULSON, GEORGE W.** Failure of ambulation in Down's syndrome. *Clinical Pediatrics*, 10(5): 265-267, 1971.

Spinal column disease, vascular accidents, environmental and psychologic mechanisms, seizures, and other neurologic disturbances are discussed as potential causes for the loss of ambulation in Down's syndrome (mongolism). Ss were 88 patients in 2 institutions for the mentally retarded, who had previously shown chromosomal and clinical diagnosis of Down's syndrome. Progressive neuromuscular degeneration was noticed in one S (CA 25 yrs), and although cranial nerve functions were intact and sensory examination was normal, roentgenograms of the cervical area showed a chronic dislocation. Since mongoloid children have a high incidence of hypotonia and lax ligaments, it is possible that cord compression may occur suddenly; incomplete fusion of the vertebral canal may also be a factor in loss of ambulation. Another S (CA 12 yrs) became severely hemiparetic following a fever. Arteriography performed 1 month later revealed occlusion of the middle cerebral artery. In Down's syndrome, acute hemiplegia may be caused by other sources, but vascular occlusion is the most probable cause. A third subject who had been in an institution since age 7 and had learned to walk, regressed slowly to a bedridden state when a favorite matron left the institution. Minor changes in the environment of a child who is late in walking and low in intelligence may be a major cause for reduction or cessation of ambulation. Loss of ambulation may also be caused by seizures, as a result of abnormal tissue in brain nodules, or by abnormalities of bone development, such as hip underdevelopment and dislocation. (8 refs.) - *T. C. Turner.*

Ohio State University Hospital
Columbus, Ohio 43210

2954 **SHAPIRO, LAWRENCE R.; HSU, L.Y.F.; & HIRSCHHORN, KURT.** Extra posterior cervical skin. *Journal of Pediatrics*, 78(4): 722, 1971. (Letter)

The suggestion that Turner's syndrome should be included in the list of chromosomal aberrations associated with excess posterior cervical skin without webbing was advanced because of a recent report of chromosomal abnormalities associated with this physical finding. One of these aberrations was XO/XY mosaicism (45,X/45,XY). A nonchromosomal disorder (cerebrohepatorenal syndrome) was also observed to be associated with the excess posterior cervical skin. The report was for the purpose of suggesting that this physical finding in an infant may be of value in raising suspicion of a nonspecified chromosomal aberration. Other cases of chromosomal aberrations associated with extra posterior cervical skin are expected to be observed in the future. (1 ref.) - *M. S. Fish.*

Mt. Sinai School of Medicine
New York, New York

2955 **BEAN, S. CHARLES; & LUSSER, MARTHA A.** Extra posterior cervical skin. *Journal of Pediatrics*, 78(4): 722, 1971. (Letter)

A recent report of 3 infants with excess posterior cervical skin without webbing and with previously undescribed chromosomal abnormalities did not mention Turner's (XO) syndrome. Another case referred because of progressive edema of the dorsum of the hands and feet was found to have excess posterior cervical skin, a shield chest, decreased femoral pulses, and 45 chromosomes and XO karyotype. Since excess posterior cervical skin without webbing has been reported previously for Turner's syndrome, the

suggestion is made that this syndrome be included in the list of chromosomal aberrations associated with this physical finding. (2 refs.) *M. S. Fish.*

Children's Hospital Medical Center
Boston, Massachusetts 02155

2956 CARREL, ROBERT E.; SPARKES, ROBERT S.; & WRIGHT, STANLEY W. Partial F trisomy associated with familial F/13 translocation detected and identified by parental chromosome studies. *Journal of Pediatrics,* 78(4): 664-672, 1971.

Examination of parental chromosomes may be helpful in detecting hidden or small chromosome changes in an affected child. Initial interpretations of chromosome analyses of 3 siblings with MR and multiple congenital defects were normal; however, investigation of the mother showed her to be a balanced carrier of an F/13 chromosome translocation, and a similar finding was made for the maternal grandfather. The sister of the mother and one of the mother's daughters (a twin of one of the probands) had appearances similar to the affected sibs. Four other pregnancies of the mother resulted in spontaneous abortions. Reanalysis of the chromosomes of the 3 Ss revealed the presence of a partial F trisomy. The congenital defects of the affected Ss included multiple craniofacial abnormalities (especially prominent cheeks), congenital heart lesions, hypotonicity, and increased digital arches. Laboratory analyses of blood and urine gave normal values. (19 refs.) - *M. S. Fish.*

Tri-Counties Regional Center
Santa Barbara, California 93104

2957 HECHT, FREDERICK. Family chromosome studies. *Journal of Pediatrics,* 78(4): 735-736, 1971. (Editorial)

A recent paper has pointed out the importance of family chromosome studies in the delineation of certain clinical syndromes. The report indicated that initial chromosome studies of 3 siblings with MR and multiple congenital anomalies were interpreted as normal; however, after the mother was found to have a balanced F/13 chromosome translocation, reanalysis of the childrens' karyotype revealed the presence of partial F trisomy and that the mother had received the reciprocal translocation in balanced form from her father. In a similar study a mother had 2 children with MR and multiple malformations from different marriages. Again, initial interpretation of the chromosome studies of the children was normal; however, when metaphases from the mother and her father showed a normal number of 46 with a missing C chromosome (determined to be a C/C reciprocal translocation), the 2 affected children were shown to have received only 1 of the 2 chromosomes involved in the translocation, giving them a partial deletion and a partial trisomy for C chromosomes, most likely chromosomes 8 and 10. Physical and laboratory examinations of both parents and children may be as essential as family history in pediatric evaluation. (2 refs.) - *M. S. Fish.*

University of Oregon Medical School
Portland, Oregon 97201

2958 Controlling chromosome loss. *Nature,* 230(5293): 351, 1971.

Recent advances in experimental methods have opened up new approaches to the study of human genetics. In addition to the classical method of establishing human gene linkage groups and assigning genes to particular chromosomes by tracing familial inheritance of a particular trait, enzyme, or disease, the technique of inducing cell fusion with inactivated Sendai virus has opened up the field as an experimental science. Mouse-human hybrids of this fusion preferentially shed their human chromosomes allowing the scientist to assign to chromosomes the human genes which specify human proteins distinguishable from the murine counterparts. Although the method has restrictions, which include the observations that hybrids with human cells form only with mouse and Chinese hamster cells and the fact that distinguishing of the human from the animal proteins is often not feasible, recent findings indicate the possibility of using prefusion irradiated cells to dictate which parental set of chromosomes will be shed from a hybrid. Preferential loss has also been shown to be influenced by incorporating bromodeoxyuridine into the DNA of Chinese hamster cells, exposing them to visible light (which induces a break in the DNA strands), followed by fusion to murine cells. Preferential loss of the hamster chromosomes from the hybrid then ensues. (No refs.) - *M. S. Fish.*

2959 **CARR, D. G.** Chromosome studies in selected spontaneous abortions. III. Early pregnancy loss. *Obstetrics and Gynecology*, 37(5): 750-754, 1971.

The mean incidence of chromosomal anomalies in abortuses appears to be approximately the same (38%) throughout the first trimester. Chromosomal analyses performed on 21 abortuses with a menstrual age of 60 days or less and compared with another series of 8 specimens of the same age range disclosed that 11 of the total 29 had chromosomal anomalies and that this frequency was comparable to that obtained for another group of abortuses in the menstrual age range of 61-90 days. Previous studies have shown that in the menstrual age range of 91-120 days the frequency of chromosomal anomalies is less than 25% and drops to about 3% thereafter. Problems associated with studies of this type include lack of sufficient specimens, since few women with pregnancy loss ever reach the hospital, and the difficulty of determining the age of the abortus because of inaccurate reporting of menstrual dates and the lack of coital histories. (11 refs.) - *M. S. Fish.*

McMaster University
Hamilton, Ontario, Canada

2960 **MILLER, ORLANDO J.; WARBURTON, DOROTHY; & BREG, W. ROY.** Partial deletion of short arm of chromosome No. 4. *Journal of Pediatrics*, 78(5): 911-912, 1971. (Letter)

In response to an inquiry regarding possible difficulty in the differential diagnosis of the Smith-Lemli-Opitz syndrome from the 4p- syndrome, the resemblance of the former disorder to trisomy 13 is pointed out; however, the authors have never made a diagnosis of the Smith-Lemli-Opitz syndrome and, as a consequence, could not have made such a misdiagnosis. Examination of the facies of 13 published cases of this syndrome indicates only 1 possible case where the diagnosis might have been 4p- instead. The explanation of similarities between likely malformation syndromes does not require proposing possible undetected chromosome rearrangements, although the recent development of the quinacrine fluorescence method for chromosome identification may make the detection of such rearrangements possible. (4 refs.) - *M. S. Fish.*

Columbia University College of
Physicians and Surgeons
New York, New York 10032

2961 **MACE, JOHN W.; & CUNNINGHAM, M. D.** Partial deletion of short arm of chromosome No. 4. *Journal of Pediatrics*, 78(5): 911, 1971. (Letter)

In accordance with a recent report the similarities of clinical features of syndromes of partial deletion of the short arms of chromosomes 4 (4p-) and 5 (5p-) are discussed. The exception appears to be the cat-like cry characteristic of the 5p- syndrome. Attention is called to the similarity of the 4p- syndrome to the Smith-Lemli-Opitz syndrome. All 3 syndromes have in common the features of: hypertelorism, antimongoloid slant of the eyes, narrow ear canals, hypotonia, growth and psychomotor retardation, microcephaly, broad base of nose, abnormal ears, micrognathia, and foot deformities. Other deformities are more characteristic of one of these syndromes compared to another; however, they often are not present and may have limited usefulness in differential diagnosis when karyotypic analyses are not available. As a consequence, the question arises as to the possibility that patients considered to have the Smith-Lemli-Opitz syndrome may actually represent the 4p- patients, and as to whether other investigators have found difficulty in clinically separating these 2 syndromes. (3 refs.) - *M. S. Fish.*

University of Colorado Medical Center
Denver, Colorado 80220

2962 **WERTELECKI, WLADIMIR; & GERALD, PARK S.** Clinical and chromosomal studies of the 18q-syndrome. *Journal of Pediatrics*, 78(1): 44-52, 1971.

A study of patients with partial deletion of the long arm of chromosome No. 18 (18q-) showed abnormal immunoglobulins in 2 of the 5 patients examined; however, no conclusion regarding either the pathogenesis or the significance of this change, also reported by another investigator, could be drawn. The study population was 6 Ss who exhibited features characteristic of the 18q- syndrome: all 6 were MR (severe in 5), had characteristic facies (deep-set eyes, carp-shaped mouth), prominent antihelix and/or antitragus,

generalized hypotonia, incoordination, proximally implanted or short thumbs, and an increased number of whorls on the fingers. Short stature, hearing loss, ocular and genital anomalies, and congenital heart disease were also observed for some of the Ss. Diagnosis of 3 of the Ss was based on these characteristic clinical signs before chromosomal analysis provided confirmation. The mechanism leading to chromosomal deletion was varied; familial pericentric inversion of No. 18 chromosome occurred in one family, a rare finding where unbalanced progeny are produced. The 2 observed abnormal immunoglobulins were absence of detectable IgA in one and low level of the component in the other. (32 refs.) - *M. S. Fish.*

Medical University of South Carolina
Charleston, South Carolina 29401

2963 **KNIGHT, LOUISE A.; SAKAGUCHI, SUMIKO; & *LUZZATTI, LUIGI.** Unusual mechanism of transmission of a maternal chromosome translocation. *American Journal of Diseases of Children,* 121(2): 162-167, 1971.

A case of a partial trisomy of a B-group chromosome in a child whose mother is a heterozygote carrier for a reciprocal 5/14 translocation between a B and a D group chromosome cannot be interpreted on the basis of usual meiotic events in the mother. The proband, a 2½-year-old girl, was the product of the sixth pregnancy of a mother who had a total of 7 pregnancies and 5 miscarriages. The proband's only surviving sib was a normal healthy female. The proband had diaphragmatic hernia at birth, and tetralogy of Fallot was diagnosed at 2 weeks. At 2½ years the S was markedly cyanotic, severely retarded, had bilateral epicanthal folds, ptosis of the eyelids, a high arched palate, low-set ears, marked scoliosis, and severe muscular spasticity. She died at 4 years of age in a hospital for the MR. Chromosomal examination of peripheral lymphocytes from the proband and her family showed that her sister, father, and maternal grandmother had normal karyotypes, as did two half-siblings of the mother. The maternal grandfather was unavailable for study. The long B-group chromosome (about one-third longer than the other 3) in the proband was not observed in the mother whose karyotype was interpreted as a reciprocal 5/14 translocation, t(14+; 5q-). One explanation involves meiotic pairing of the maternal translocation chromosomes in a quadriradial configuration and 2 breaks to form the long B-group chromosome of the proband. (13 refs.) - *M. S. Fish.*

*Stanford University School of Medicine
Stanford, California 94305

2964 **HSIA, DAVID YI-YUNG; JUSTICE, PARVIN; SMITH, GEORGE F.; & DOWBEN, ROBERT M.** Down's syndrome. A critical review of the biochemical and immunological data. *American Journal of Diseases of Children,* 121(2): 153-161, 1971.

A review of data concerning enzyme changes, protein synthesis, and immunological mechanisms in Down's syndrome is presented. The principal biochemical alterations in this disorder are the increase of several enzymes in the leukocytes and of 2 in the erythrocytes with no significant changes in other tissues. Changes in leukocytes suggest that chromosome 21 may be involved in the regulation of protein synthesis in leukocytes. Studies of DNA and RNA production following phytohemagglutinin stimulation of lymphocytes indicate that excessive enzyme activity among trisomy 21 patients does not result in an increase of protein synthesis. This observation resulted from the examination of the results obtained from investigating 2 systems involving protein synthesis: in 1 system the patients appear to have impaired DNA response to phytohemagglutinin stimulation; in the other, a similar lack of RNA response occurred. The currently most promising area for the study of immunologic mechanisms in Down's syndrome derives from observations which relate the Australian antigen to this disorder. In several inst the frequency of the antigen is 28% among Down's syndrome patients, compared to 3% among other inst MR. The antigen has also been demonstrated to be related to infectious hepatitis and has been found to be common in leukemia. (64 refs.) - *M. S. Fish.*

Loyola University
Stritch School of Medicine
Maywood, Illinois 60153

2965 **KAUFMAN, ROBERT L.; RIMOIN, DAVID L.; WOOLF, RALPH B.; WARREN, RICHARD; & SLY, WILLIAM S.** Sex chromosome mosaicism (XO/XX/

XXY/XY) in a phenotypic female. *Journal of the American Medical Association*, 215(12): 1941-1944, 1971.

A tall, 26-year-old woman of normal female phenotype with secondary amenorrhea and infertility was found to have four stem-line sex chromosome mosaicism (XO/XX/XXY/XY) in lymphocyte and skin cultures. A pneumogynogram suggested an enlarged left gonad and an atrophic right gonad. At laparotomy a total hysterectomy and bilateral salpingogonadectomy were performed. A left adnexal cyst showed hilar cell hyperplasia, and the right adnexal "streak" showed normal mesonephric remnants. Surgery was mandatory in this case because of the high probability of malignancy in Y-containing cell lines in abdominal gonads. (12 refs.) - *E. L. Rowan.*

500 S. Kingshighway Blvd.
St. Louis, Missouri 63110

2966 Environmental factors in Down's syndrome. *Lancet*, 1(7711): 1223-1224, 1971.

The incidence of Down's syndrome is approximately 1 in 636 births. Chromosome studies have revealed that the majority of individuals with Down's syndrome are trisomic for chromosome 21. About 4 to 5% of those with Down's syndrome are trisomic for chromosome 21 in association with translocation. Mosaic individuals have 1 cell line which is trisomic for chromosome 21. Among the environmental factors with a suspected relationship to Down's syndrome are maternal fluoroscopic and therapeutic radiation, virus infections, and maternal thyroid antibodies. Present evidence supports the theory that the birth of a mongoloid child is due to an error of gametogenesis in the mother. Future research will seek to identify the parent and the stage of gametogenesis at which the error related to trisomy 21 occurs. (19 refs.) - *J. K. Wyatt.*

2967 **MILLER, D. A.; ALLDERDICE, P. W.; MILLER, O. J.; & BREG, W. R.** Quinacrine fluorescence patterns of human D group chromosomes. *Nature*, 232(5305): 24-27, 1971.

The use of quinacrine fluorescence patterns to identify 10 different abnormalities of chromosomes 13, 14, and 15 appears to be associated with advantages over autoradiographic and morphological methods. In studies performed on 18 Ss with D group chromosome abnormalities, autoradiographic findings agreed for 8 of the 9 cases compared. Morphology is inadequate in distinguishing the 3 pairs of homologous group D chromosomes because of small differences in length and arm ratio, while the more accurate autoradiographic analysis is limited in usefulness because it is time consuming, and errors in identification can occur. Quinacrine fluorescence patterns provide the best method so far available and allow rapid identification of both specific chromosomes and of normal and translocated chromosomes. Chromosome 13 is more easily distinguished by fluorescence than are chromosomes 14 and 15. In the present study fluorescence patterns were clearly visible for 1 t(13q13q), 2 t(13q14q), and 6 t(14q21q) cases of translocation. The technique also was found useful in the study of 2 cases of trisomy 13 syndrome and in cases of 13q-, 14p-, and 15p- deletion. In a case where a patient had 7 D group chromosomes but did not have clinical features of trisomy 13 syndrome, the mother was found to have a reciprocal translocation involving chromosomes 9 and 14, and the son was found to be trisomic for most of chromosome 14. (18 refs.) - *M. S. Fish.*

Columbia University
College of Physicians and Surgeons
New York, New York 10032

2968 **BENSON, PHILLIP F.; & SOUTHGATE, JENNIFER.** Diminished activity of platelet monoamine oxidase in Down's syndrome. *American Journal of Human Genetics*, 23(2): 211-214, 1971.

Monoamine oxidase (MAO) activity in platelets from children with Down's syndrome (DS) was found to be significantly lower than that observed in platelets from matched controls, suggesting that the low serotonin (5-hydroxytryptamine, 5HT) level in DS is due to a mechanism other than an enhanced rate of 5-HT metabolism. Platelets obtained from blood samples of 22 children with DS confirmed by karyotypic analysis and from samples of 22 controls matched for age, domicile, and sex were isolated by centrifugation, suspended in water, and analyzed spectrophotofluorometrically for MAS activity. Results showed that the mean

MAO activity (expressed as grams of 4-hydroxyquinoline formed per mg platelet protein per 30 min) of the DS patients was 0.81 units, S. D., 0.38, compared to a value of 1.24 units, S. D., 0.70 for controls (p=0.01-0.02). Possible explanations for both low 5HT levels and low MAO activity in DS may be that enzymes catalyzing the 5-hydroxyindole pathway are repressed due to a relative deficiency of tryptophan or that the trisomic chromosome 21 may carry a regulatory gene for MAO. (21 refs.) - *M. S. Fish.*

Guy's Hospital Medical School
London, S. E. 1, England

2969 **WILKINS, ROBERT H.; & BRODY, IRWIN A.** Down's syndrome. *Archives of Neurology*, 25(1): 88, 1971.

As a superintendent of an asylum for idiots Dr. John Langdon Haydon Down observed a group of patients with characteristic features, such as prominent epicanthal folds, which suggested a mongol appearance. Likely influenced by current theories on evolution, Down apparently viewed this type of idiocy as reversion to an earlier phylogenetic type, possibly due to parental tuberculosis, and proposed an ethnic classification for the disorder. Since the syndrome is now known to have a genetic basis (47 chromosomes with trisomy of a G group chromosome) and to occur in a variety of races, the suggestion is advanced that Down be honored for his observations in this field by having his name employed as the sole designation of this syndrome. (9 refs.) - *M. S. Fish.*

Duke University Medical Center
Durham, North Carolina 17706

2970 **JENSEN, ROBERT D.; & MILLER, ROBERT W.** Retinoblastoma: Epidemiologic characteristics. *New England Journal of Medicine*, 285(6): 307-311, 1971.

A nationwide study was made of 269 death certificates of persons under 15 years of age who died of retinoblastoma during the period 1960 to 1967 in the United States (mortality series) and of 1,623 hospital records of children with the disease (hospital series). Comparison of mortality rates for whites and nonwhites by single year of age revealed peaks in mortality for both groups at 2 to 3 years of age, but the peak for nonwhites was 2.5 times greater than that for whites, a finding that is unique among child cancers. In the hospital series, 53 of the 1,077 children with adequate information concerning physical findings manifested major congenital malformations, with the incidence of SMR individuals significantly exceeding expectation. In 39 other children in the hospital series, retinoblastoma occurred with malformations which were felt to be transmitted genetically. The findings indicate that MR with retinoblastoma may represent a variant of the D-deletion syndrome, and children in this category merit cytogenetic study. Thirty children had second primary cancers that appeared to be radiation induced. In 11 other children, second primary cancers were not radiogenic. The occurrence of osteosarcoma of the femur in 3 cases suggests an innate tendency to this cancer among children with retinoblastoma. The peak in the age at diagnosis of retinoblastoma, occurring so soon after birth, indicates that the neoplasm has its origins prenatally, though not necessarily prezygotically. (28 refs.) - *B. J. Grylack.*

National Cancer Institute
Bethesda, Maryland 20014

2971 **PARRINGTON, JENNIFER M.; & EDWARDS, JOHN H.** Patau's syndrome with D_1 duplication-deficiency derived from a maternal D group pericentric inversion. *Annals of Human Genetics*, 35(1): 35-45, 1971.

A newborn male infant presented with marked congenital anomalies and a clinical diagnosis of trisomy D that was consistent with Patau's syndrome. The propositus was the product of a sixth pregnancy and was born when the mother was 31 and the father 44. He had 2 healthy male siblings. Two previous pregnancies had miscarried at 3 months, and 1 infant was stillborn with generalized abnormalities. In skin and blood chromosome samples from the S the chromosome number was 46, but there were only 5 normal members of the D group. An extra chromosome in the C group(46XY Dp^+) was thought to resemble 1 of the larger members of the C group (6 to 7X). In the mother the chromosome number was 46, but in place of a D

group there was an unpaired, nonsatellited submetacentric chromosome which was the same length as a D-group chromosome. The father and 2 healthy sons had normal male karyotypes. It appears that the marker chromosome in the mother may be a D-group pericentric inversion and that the abnormal D group in the S is the duplication-deficiency product of crossing over within the loop formed by the inverted chromosome and its homologue during meiosis. (28 refs.) - *B. J. Grylack.*

University College
London, England

2972 **PENROSE, L. S.; & LOESCH, DANUTA.** Dermatoglyphic patterns and clinical diagnosis by discriminant function. *Annals of Human Genetics*, 35(1): 51-60, 1971.

Differences between normal karyotypes and trisomies D, E, G, and 4-5 shorter arm, or Bp, deletion were established by combining data from previous observations so as to construct 4 sets of discriminant weightings. With the use of characters on the fingers, palms, and soles, an attempt was made to improve determination of dermatoglyphic patterns and clinical diagnosis by discriminant function. Two examples of the use of the discriminants were presented. The first consisted of an unknown diagnosis suspected of mongolism, and the second was a known mosaic female mongol. During calculation of a discriminant value for any single case, the presence of a pattern was scored in the form of a percentage frequency for each pair of corresponding fingers, palms, and soles. The use of discriminant function in comparing abnormal groups can also provide information on the interrelation between trisomies and monosomies, between the various types of sex chromosomal anomalies, or on the relationships of mosaics and partial trisomies to the corresponding complete syndromes. (12 refs.) - *B. J. Grylack.*

Harperbury Hospital
Nr. St. Albans, Hertfordshire, England

2973 **LAMBOTTE, C.; KEUTGEN, H.; & FREDERIC, J.** Le genotype 47,XYY (The 47,XYY genotype). *Acta Paediatrica Belgica*, 25(3): 154-183, 1971.

An increased incidence of XYY genotypes has been found among tall male prisoners and other institutionalized males. While intelligence is most frequently affected in chromosomal aberrations, it is questionable whether XYY individuals are mentally subnormal. XYY males have been shown to be aggressive and to tend towards sexual offenses and homosexuality. Accessory signs include neurological difficulties, osteoarticulary malformations, anomalies of the genital organs, and dermatoglyphic anomalies. Genital malformations, skeletal anomalies, and psychomotor retardation prevail with great variation in XYY children. The frequency of the gonosomic XYY in the white population is probably higher than 0.1% and similar to that of the Klinefelter XXY syndrome. The overwhelming majority of XYY children and adults are well integrated into the normal population, from which they can be distinguished only by a greater than normal average height and often by the presence of some impulsiveness appearing during childhood or adolescence. The pediatrician should thus be aware of the possibility of an XYY chromosomal constitution in children who manifest these traits. It remains for future endocrinologic studies to evaluate the effects of the extra Y chromosome on the secretion and perhaps even the structure of certain endocrine glands and for anthropologic, dermatoglyphic, and biologic research to produce an easily applied test for the detection of XYY children. (115 refs.) - *B. J. Grylack.*

Hopital de Baviere
B-4000 Liege, Belgium

2974 **LAMBOTTE, C.; FREDERIC, J.; STAINIER, L.; JOVANOVIC, M.; & BAUDUIN, P.** Premier cas de deletion au niveau du bras long d'un chromosome 5: Caryotype 46, XY/46,XY,5q (First case of deletion at the level of the long arm of a chromosome number 5: Karyotype 46,XY/46,XY,5q). *Acta Paediatrica Belgica*, 25(3): 143-153, 1971.

A male infant presented with a series of minor dysmorphias and complex cardiovascular malformations consisting of Fallot's pentalogy and persistence of a left superior vena cava ending in the left atrium. The patient manifested a chromosomal mosaicism, half of the cells being normal and the others being characterized by a deletion of approximately 20% at the level of the long

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arm of a chromosome number 5. Karyotypic examination of the parents did not reveal any anomaly, and the chromosomal deviation of the patient was attributed to random occurrence. No similar chromosomal aberration has been recorded in the literature. (3 refs.) - *B. J. Grylack.*

Hopital de Baviere
B-4000 Liege, Belgium

2975 **LAMBOTTE, C.; CARLIER, G.; FREDERIC, J.; & KEUTGEN, J.** Presentation d'un cas de translocation C/G. Caryotype 46,XY,t(Cq-;Gq+); parents 46,XX et 46,XY (Presentation of a case of C/G translocation. Karyotype 46,XY,t(Cq-;Gq+); parents 46,XX and 46,XY). *Acta Paediatrica Belgica,* (25(2): 119-126, 1971.

A hypotrophic 5-year-old male with severe psychomotor retardation and dysmorphias of the facies and ears presented with an apparently balanced translocation between a chromosome of group C (probably an 11) and a chromosome of group G (probably a 22). Chromosomal examination of the parents did not reveal any anomaly. The most logical explanation for the rare association of a balanced translocation and an abnormal phenotype in the patient is that a small fragment of the chromosome was eliminated in the course of the translocation and that this loss caused the manifested anomalies. It is possible, however, that no phenotypic consequences followed the translocation and that the association between the visible chromosomal aberration and the presented anomalies was due to random occurrence. (6 refs.) - *B. J. Grylack.*

Universite de Liege
B-4000 Liege, Belgium

2976 **RIDLER, M. A. C.** Banding patterns of metaphase chromosomes in Down's syndrome. *Lancet,* 2(7720): 354-356, 1971.

A simple and inexpensive new Giemsa staining technique for the investigation of human metaphase chromosomes has been applied to the differentiation of the G-group chromosomes and the classification of the extra chromosome in cultured lymphocytes of patients with Down's syndrome. In preparations from 5 male Ss, 80 selected cells were examined. Denaturation followed by Giemsa staining revealed differences in the banding patterns of the small chromosomes only in the best cells, which consistently showed 2 distinct classes of chromosomes in the G group. A larger pair showed a prominent darkly stained centrometric area with the occasional occurrence of 1 or 2 less dense regions in the long arms. The remaining 3 chromosomes were usually smaller and more densely stained. Y chromosomes were distinguished by their characteristic shape, dark staining, and sometimes by 3 bands in the long arm. Down's syndrome appears to involve the more densely stained chromosomes of the G group. (7 refs.) - *B. J. Grylack.*

Harperbury Hospital
Nr. St. Albans, Hertfordshire, England

2977 **FEDRICK, JEAN.** Down's syndrome: Sex difference in relation to maternal age. *Lancet,* 2(7714): 45, 1971. (Letter)

Data from the 1958 British perinatal mortality survey were analyzed to compare the mean maternal age of males and females with Down's syndrome. Results showed that mean maternal age of male mongols was slightly higher than that of the females and the reverse was true for stillbirths and neonatal deaths. It seems that sex has little bearing on the etiology of mongolism. (7 refs.) - *T. C. Turner.*

University College
Department of Human Genetics and Biometry
London, England

2978 **WALKER, S.; GREGSON, N. M.; & HIBBARD, B. M.** Nuclear sex and antenatal diagnosis of Klinefelter's syndrome. *Lancet,* 2(7721): 430, 1971. (Letter)

The sex of the fetus was investigated in 50 specimens of amniotic fluid (14 weeks gestation to almost full-term), using sex-chromatin study and quinacrine fluorescence. Scoring for sex chromatin should include only nuclei which are evenly stained and have a regular outline, and at least 100 cells should be scored. Of the 50 samples, 31 gave a score of 0-2% chromatin-

positive and a fluorescence score of 40-84%, with sex diagnosed as male. Eighteen gave a score of 15-37% chromatin-positive and 0-14% fluorescence-positive, with sex diagnosed as female. Sex was confirmed in all cases. In 1 case, scores of 34% chromatin positive and 74% fluorescence positive were recorded, and Klinefelter's syndrome was suspected. At birth chromosome counts of 2n=47 were made. Autoradiography showed a late-labeling chromosome present in Group C, and a buccal smear yielded a score of 34% sex-chromatin positive, confirming Klinefelter's syndrome. Delivery was normal and no physical abnormalities were noted. Use of either technique alone (sex chromatin or fluorescence) would have led to an error in the diagnosis of chromosomal sex. (2 refs.) - *K. L. Woodworth.*

University of Liverpool
Liverpool L69 3BX, England

2979 **BAIKIE, A. G.** Banding patterns of metaphase chromosomes in Down's syndrome. *Lancet,* 2(7722): 494, 1971. (Letter)

The mongol chromosome in Down's syndrome has been variously identified as a member of the later replicating pair of Group G (with the Ph apparently derived from the early replicating pair), or belonging to the shorter pair (and the Ph derived from the longer pair). The mongol chromosome has been referred to as no. 21 while others attribute it to trisomy 22. Complicating it further, trisomy-21 has been used as a synonym for Down's syndrome, and trisomy-22 to denote a rarer condition. The differences must be resolved by international agreement. (4 refs.) - *B. Berman.*

Radcliffe Infirmary
Oxford, England

2980 **THELEN, THOMAS H.; ABRAMS, DONALD J.; & FISCH, ROBERT O.** Multiple abnormalities due to possible genetic inactivation in an X/autosome translocation. *American Journal of Human Genetics,* 23(4): 410-418, 1971.

A female child with multiple congenital anomalies and a translocation that involved the X chromosome was studied with interest because her abnormalities appeared to be due to genetic inactivation rather than to a loss or gain of genetic material. The S had evidence of delayed mental and motor development along with congenital somatic anomalies when seen at 26 months of age. All of the peripheral blood cells examined lacked a C group and a number 18 chromosome but had an additional large metacentric chromosome and a small metacentric marker. From both the clinical and cytogenetic findings, the karyotype of the patient was interpreted as the result of a reciprocal translocation involving the interchange of a large segment of the long arm of a number 18 chromosome with the small segment of the short arm of an X chromosome [46,Xt(Xp+;18q−)]. The prognosis for the S is poor, although it may be somewhat better than that for a patient with a deletion of the long arm of chromosome 18, since the translocated autosomal segment is not inactivated, at least in the cells with an inactive normal X. (33 refs.) - *B. J. Grylack.*

Central Washington State College
Ellensburg, Washington 98926

2981 **HECHT, FREDERICK; & KIMBERLING, WILLIAM J.** Patterns of D chromosome involvement in human (DqDq) and (DqGq) Robertsonian rearrangements. *American Journal of Human Genetics,* 23(4): 361-367, 1971.

Lymphocytes from 19 unrelated persons, including 4 with t(DqDq) and 15 with t(DqGq) translocations, were studied in an attempt to delineate the overall patterns of composition of Robertsonian translocations. Autoradiographic analysis showed that the (DqDq) rearrangements were (13q14q) in 3 cases and (14q15q) in 1 case. The (DqGq) translocations were (14qGq) in 11 cases, (15q21q) in 3 cases, and (14qGq) in 1 case where the nature of the G chromosome was not known. Based upon the data from this study and the literature, the pattern of involvement in (DqDq), excluding the case of (15q15q), which could be an isochromosome, is nonrandom (x^2=37.1, $p<.001$, 2 df). It also is very highly nonrandom in (Dq21q) (x^2=9, 224, $p<.001$, 2 df). Due to evidence that Robertsonian exchanges in man are extraordinarily common in comparison with other types of translocations and that they usually appear to be of spontaneous origin, it is suggested that Robertsonian chromosomes

may result from an orderly, nonrandom process such as meiotic pairing and exchange, rather than from random breakage and fusion. (45 refs.) - *B. J. Grylack.*

University of Oregon Medical School
Portland, Oregon 97201

2982 **DUCOS, J.; MARTY, YVONNE; SANGER, RUTH; & RACE, R. R.** X_g and X chromosome inactivation. *Lancet,* 2(7717): 219-220, 1971. (Letter)

Although Lynn's theory of random inactivation of one or the other X chromosome of a normal mammalian female is now almost universally accepted, a remaining question is whether inactivation involves all the loci in the human X. Evidence suggests the X_g locus in X-linked X_g blood groups is not involved when carried on a structurally normal X and that human X inactivation is not complete. Recent discovery of a twin chimera pair with a mixture of group-O and group-A_1B cells also suggests inactivation of a structurally normal human X is not complete. (7 refs.) - *B. Berman.*

Hopital Purpan
Toulouse, France

2983 **SHAPIRO, LAWRENCE R.; REDNER, WALLACE J.; & INNELA, FILOMENA P.** Australia antigen and Down's syndrome. *Lancet,* 2(7717): 215-216, 1971. (Letter)

A comparison of 50 Ss with trisomy-21 Down's syndrome and 2 females with translocation Down's syndrome (G/G and D/G) with 17 cytogenetically normal MRs revealed Australia antigen (Au) in 9 Down's Ss and 2 controls, with an anticipated decrease of incidence in females. The 2 translocation Ss (both females) had no detectable Au. Speculation was indicated on a karyotype relationship between Down's syndrome and Au. (4 refs.) - *B. Berman.*

Cytogenetics and Clinical Laboratories
Thiells, New York

2984 **PETRE-QUADRENS, O.; & DE GREEF, A.** Effects of 5-HTP on sleep in mongol children: Preliminary results. *Journal of the Neurological Sciences,* 13(1): 115-119, 1971.

5-hydroxytryptophan (5-HTP) increased eye movement density in mongol children during sleep. The density of eye movements during rapid eye movement sleep (REM) in normal and mongol neonates of similar gestational age was compared. The number of eye movements during sleep was decreased in the mongols, and the distribution of the high density eye movement patterns within the consecutive REM epochs differed considerably from those seen in normal newborn infants. After 5-HPT intake, the density of the eye movements increased in mongol children older than 1 year of age. In addition, eye movement density distribution during sleep showed a periodic alternation of high and low density REMs similar to the REM rhythm seen in normal children. (10 refs.) - *S. Glinsky.*

Born-Bunge Foundation for Scientific Research
Bercham-Antwerp, Belgium

2985 **FERDON, NONA M.** Chromosomal abnormalities and antisocial behavior. *Journal of Genetic Psychology,* 118 (Second half): 281-292, 1971.

Studies seem to indicate a definite correlation between occurrence of X or Y chromosomal aberrations and aberrant behavior, although much of the evidence is derived from institutionalized populations. Almost all research in sex-chromatin positive men has shown this association. A 1965 study found 7 XYY and 1 XXYY males in 197 prison inmates. Casey found 12 XYYs in 50 mentally subnormal men. Hunter found 3 XYYs in 29 Ss, with IQs of 77, 78, and 91. Other research has continued to show this association. Male XXYs are distinguished from normals by various physical abnormalities; there are no comparable abnormalities among XYY males. More longitudinal studies are needed for definite conclusions on an association between superfluous X or Y chromosomes and violent antisocial behavior. (51 refs.) - *B. Berman.*

University of Hawaii
Honolulu, Hawaii

2986 **ABRAMS, NAOMI; & PERGAMENT, EUGENE.** Childhood psychosis combined with XYY abnormalities. *Journal of*

Genetic Psychology, 118 (First half): 13-16, 1971.

A history is presented of one of 3 cases of XYY complement and psychosis. S was first seen at age 22 months, unable to speak and apparently severely retarded intellectually, with inconsistent response to sound. Cytogenetic analysis revealed aneuploidy with a modal chromosome number of 47 (the extra chromosome was a small acrocentric, and 2 chromosomes had a Y morphology). The axial triradius of both palms was somewhat displaced distally and the digital pattern showed 5 arches. S showed delayed motor development, high arched palate, and hypertelorism. There was no attempt to use sound for communication, and his behavior was described as psychotic. Questions of infantile autism, Down's syndrome, and biochemical, physiologic, and genetic roles need resolving in this case. (11 refs.) - *B. Berman.*

Michael Reese Hospital and Medical Center
Chicago, Illinois

2987 PERRY, TRACY B. Down's syndrome: Sex difference in relation to maternal age. *Lancet*, 2(7718): 263, 1971. (Letter)

A summary of available data indicates that sex difference per se probably is not an important factor to be considered in studies of maternal age and mongolism. Although certain studies (Largey and Largey, Spencer) seem to indicate significant differences in mean maternal age (MMA) for male and female mongols, their findings are not confirmed by other studies. A summary of data from all these studies (1,384 Ss) shows a MMA of 35.73 yrs for female mongols (589 Ss) and 35.59 for males (795 Ss), a difference of only 0.14 yrs. While a test for statistical significance is not possible, this small difference certainly is not significant. It is noteworthy that the number of male mongols in the studies outnumbers the number of females by 4 to 3. An analysis of maternal-age data on 183 mongols at the Montreal Children's Hospital (1951-1971) shows a MMA for female mongols (81 Ss) of 32.77±0.81 yrs and for males (102 Ss) of 31.97±0.71 yrs. The difference of 0.80 yrs is not significant ($p=0.46$). The apparent sex differences probably are due to the smaller samples. (8 refs.) - *S. M. Rucker.*

Montreal Children's Hospital
Montreal 108, Quebec, Canada

2988 ROS, Y.; HAYEZ-DELATTE, F.; & VAN DE POEL, H. Down's syndrome: Sex difference in relation to maternal age. *Lancet*, 2(7718): 264, 1971. (Letter)

Data on 252 Ss showed that the mean maternal age (MMA) at birth of female mongols (108 Ss) was 32.94, and that of mongol males (144 Ss) was 34.39. This parallels the Largey and Largey findings (females 33.28 yrs, males 36.25 yrs; significant at $P<0.05$), but the sex difference in this study was not significant ($t=1.53$; $P>0.05$). The mean paternal age at birth of female mongols was 35.21, that of males 36.29, the difference also not being significant ($t=1.04$; $P>0.05$). (3 refs.) - *S. M. Rucker.*

Universite Libre de Bruxelles
Belgium

2989 QAZI, QUTUB H.; & LANMAN, JONATHAN T. Down's syndrome: Sex difference in relation to maternal age. *Lancet*, 2(7718): 263-264, 1971.

Data on 130 Ss with Down's syndrome (1961-1969) show that mean maternal age (MMA) at time of birth for female mongols (54 Ss) was 33.67 yrs, significantly higher ($p<0.025$) than the 30.08 yrs for male mongols (76 Ss). This conflicts with the observations of Largey and Largey, whose data show that the MMA of female mongols at birth was significantly lower than that of males. Earlier studies showed a slightly higher MMA for female mongols than for males, but a statistically significant difference was not demonstrated. Most published series indicate a higher proportion of male than female mongols; this was true of this study (1.4:1). This study also showed a significantly greater number of male than female mongols born to mothers in the 15-24 yr age group (2.7:1) than to mothers in older age groups (1.1:1). Assuming that the ratio of male to female mongols at conception is equality, this data would seem to indicate that more female than male mongols conceived by young mothers are spontaneously aborted. (4 refs.) - *S. M. Rucker.*

State University of New York
Downstate Medical Center
Brooklyn, New York

2990 **JANCU, JULIUS.** Radioulnar synostosis. *American Journal of Diseases of Children,* 122(1): 10-11, 1971.

The finding of radioulnar synostosis (RUS) in an isolated generation is highly suggestive of a sex chromosome abnormality. The relative frequency of proximal RUS in cases of sex chromosomal alterations is revealed by a review of the literature reporting Klinefelter's syndrome and its chromosomal variations (1942-1970). In the cases reported, at least 5 typical variations of karyotype have been demonstrated. In a series of 28 cases with XXXXY chromosomal karyotypes reported by Houston, 9 had RUS. Eighteen cases of sex chromosomal abnormalities taken from the literature are listed in which RUS was present. Nevertheless, the presence of RUS is not a specific diagnostic sign. (21 refs.) - *S. M. Rucker.*

Central Emek Hospital
Afula, Israel

2991 **McCORMICK, DAVID P.; MEYER, WALTER J.; & *NESBIT, MARK E.** Coexistence of Hodgkin's disease and Down's syndrome. *American Journal of Diseases of Children,* 122(1): 71-73, 1971.

A child with Down's syndrome (trisomy 21-22) developed Hodgkin's disease. As far as is known, this is the first reported case of a child with the combination, although an adult with the combination has been reported previously. The child (8 yrs old, white male) developed swelling of the left cervical lymph nodes 4 mos prior to admission. Biopsy showed many Reed-Sternberg cells, scattered eosinophils, and pleiomorphic lymphoid cells consistent with Hodgkin's disease. While the maternal grandmother died of gastric carcinoma, there was no other family history of cancer, leukemia, or chromosomal defects. The S was given radiotherapy. Two months later he developed high fevers. A laparotomy revealed metastases in the periaortic nodes and liver. Despite treatment with mechlorethamine hydrochloride and vinoblastine sulfate (Velban), S developed pancytopenia and died with progressive respiratory disease. Autopsy revealed that Hodgkin's disease was widespread. There were also cytoplasmic inclusions consistent with a diagnosis of cytomegalic inclusion disease. Further study is required to demonstrate whether there is increased incidence of solid tumors of the lymphoreticular system of Ss with Down's syndrome. (18 refs.) - *S. M. Rucker.*

*University of Minnesota Hospital
Minneapolis, Minnesota 55455

2992 **JENKINS, MELVIN E.; EISEN, JAMES; & SEQUIN, FRANCOIS.** Congenital asymmetry and diploid-triploid mosaicism. *American Journal of Diseases of Children,* 122(1): 80-84, 1971.

A case is reported in which there is an association between congenital asymmetry and diploid-triploid mosaicism (DTM). This association has been reported only twice previously, but may be more frequent than previously recognized. The S was a female (9½ yrs) with congenital asymmetry manifested by body structure larger on the right than on the left, a normal chromosome karyotype in peripheral blood lymphocytes, and diploid (2n)-triploid (3n) chromosomal mosaicism in skin fibroblasts. S was somewhat premature and considerably underweight at birth (3 lb 8 oz). A younger sibling and 3 children of the mother from a previous marriage were reportedly normal. Most laboratory findings were normal, although electroencephalogram revealed slowed activity over the left and right hemispheres and psychological examination showed moderate MR. Physical characteristics other than asymmetry included down-curved edges of the lips, incurved fifth digits, scoliosis with a convexity to the right, a soft systolic functional murmur at the third left interspace, and infantile sexual development, which suggests a variant form of Silver's syndrome. She was also of moderately short stature. There was clinodactyly with a single crease on the left fifth digit. The two previously reported cases had similar characteristics. There are 2 possible explanations of the 2n-3n mosaicism: (1) A 2n sperm might unite with a normal oocyte to form a 3n zygote, or (2) double fertilization could result in a 3n zygote. In this S, mosaicism was demonstrated repeatedly in the skin from both sides of the body. Chromosome karyotypes should be obtained from several tissues in Ss with congenital asymmetry. (37 refs.) - *S. M. Rucker.*

University of Nebraska College of Medicine
Omaha, Nebraska 68105

2993 **SINHA, ANIL K.; NORA, J. J.; & PATHAK, SEN.** Isochromosomes arising from a human 'C'-autosome. *Human Heredity*, 21(3): 231-237, 1971.

Cytological and clinical studies were done on an abnormal human female who was found to possess 2 chromosomally distinguishable cell lines. Severe growth and developmental retardation characterized the clinical course of the Negro patient, and facial and bony anomalies were observed. Sex chromatin bodies of normal size were found in 22% of the buccal mucosal cells. In 10% of the metaphase leukocytes studied, the chromosome count was 47. In these cells, a 'C'-group chromosome was missing, while 2 unequal metacentric chromosomes were present. It was also noted that 1 or the other metacentric chromosome had replaced a regular 'C'-group member in a number of cells that had 46 chromosomes. Inasmuch as the abnormal elements were unequal metacentric chromosomes and a 'C'-group autosome, a submetacentric chromosome, was lacking, it appeared that the missing autosome underwent a misdivision at its centromere which resulted in 2 unequal metacentric chromosomes, or presumptive isochromosomes. Such a trisomic state of the C-autosome seemed responsible for the phenotype features of the patient. (7 refs.) - *B. J. Grylack.*

Texas Children's Hospital
Houston, Texas 77025

2994 **LEHRKE, ROBERT; THELEN, THOMAS; & LEHRKE, ROBERT, JR.** Syndrome associated with group-D chromosome deletions. *Lancet*, 2(7715): 98-99, 1971. (Letter)

A 25-year-old PMR Caucasian male presented with a characteristic dysmorphogenetic syndrome associated with a group-D chromosome deletion. The patient is ambulatory but with rather stooped posture, does not speak, is not trained for bladder or bowel control, and has a mongoloid-like facial appearance. His cranial circumference of 53 cm is below the normal limits. Attention was first drawn to his anomalous genital development when, at age 15, he appeared to menstruate through a fistula at the base of his bifid scrotum. The urethra is at the base of a penis of relatively normal size. The pubic escutcheon is of female pattern, and slight gynecomastia contributes to the impression of intersex. On examination from leukocytes, all cells studied were 46,XY,Dr. Autoradiography revealed that the ring was formed from the late-replicating D1 (number 13) chromosome. Minimal chromosome deletion in this S may account for his relatively good physical health and long life and for the absence of hand and eye deformities. It is also possible that the group of defects reported is a consequence of severely retarded development in general, which may itself be the result of many different factors, including various forms of partial aneuploidy. (16 refs.) - *B. J. Grylack.*

Brainerd State Hospital
Brainerd, Minnesota

2995 **GRANOFF, DAN M.; & PRESTON, MALCOLM S.** Cri-du-chat syndrome—an unhelpful designation. *Lancet*, 2(7715): 99-100, 1971. (Letter)

Three female patients were described with a characteristic syndrome associated with partial deletion of the short arm of chromosome 5 (5p-). The major features of this syndrome are severe MR, microcephaly, failure to thrive, and a feeble, pleading cry present early in infancy which is probably related to neurological damage or to an abnormality of laryngeal function. Spectrograms of the cries of a patient with the 5p- syndrome and a patient with neurological damage associated with intrauterine infection from cytomegalic virus were found to exhibit higher minimum and maximum pitch than the spectrogram of the cry of a normal child. Since spectral analysis of cries produced by infants with congenital hydrocephalus and hyperbilirubinemia also shows abnormally high minimum and maximum pitch, however, it appears that high pitch in itself does not differentiate the 5p- syndrome from other entities associated with brain damage or anomalies of the trachea and larynx. In addition, the cries of cats are not uniform with regard to certain spectral properties, especially pitch. The lack of consistent correspondence between the spectral properties of cries in cats and cries in children with this condition suggests that 5p- is a more suitable name than cri-du-chat syndrome. (7 refs.) - *B. J. Grylack.*

Johns Hopkins University School of Medicine
Baltimore, Maryland 21205

2996 **COHEN, MAIMON M.; & DAVIDSON, RONALD G.** An inherited pericentric chromosomal inversion (46, inv3 [p–q$^+$]) associated with skeletal anomalies. *Journal of Pediatrics*, 79(3): 456-462, 1971.

A 3-month-old male and his mother were both afflicted with identical skeletal anomalies of the fingers and toes (syndactyly and absence of digits). The mother had no other detectable defects and seemed to be of normal intelligence. Cytogenetic studies revealed an abnormal No. 3 chromosome in both peripheral lymphocytes and fibroblasts of mother and child. Evidence from morphology, mensuration, and autoradiography supports the conclusion that the abnormal chromosome observed represents a pericentric inversion of a No. 3 chromosome (46, inv3[p-q$^+$]). This is the first reported instance of a pericentric inversion of a No. 3. (19 refs.) - *K. L. Woodworth.*

86 Hodge Avenue
Buffalo, New York 14222

2997 **NEWMAN, STEPHEN J.; MADDEN, DAVID L.; GITNICK, GARY L.; & *SEVER, JOHN L.** American Journal of Diseases of Children, 122(2): 129-133, 1971.

Agar gel double diffusion and complement fixation (CF) tests for Australia antigen and antibody were performed on serum from selected groups of patients with MR, hepatitis, leukemia, and Burkitt's lymphoma and from "normal" reformatory inmates in an effort to determine the efficacy of these methods and the frequency and role of Australia antigen in these populations and disease states. Results showed that 34.1% of 138 patients with Down's syndrome were positive for the antigen by CF testing, while 10.0% of 161 patients with MR of other etiologies were positive. Of 27 MR patients with acute infectious hepatitis acquired during an epidemic, all were negative for the antigen. However, 61.8% of 34 hospitalized patients with acute hepatitis of unknown type were antigen-positive. None of 27 serum specimens from pregnant women who delivered children with Down's syndrome was antigen-positive. There was a 79.3% concordance in all specimens between agar gel diffusion and CF. The study confirmed the strong association between Australia antigen and hepatitis, particularly "serum" hepatitis, and suggested that genetic or host factors as well as environmental causes may be responsible for the persistence of the antigen in patients with Down's syndrome. (20 refs.) - *B. J. Grylack.*

*National Institutes of Health
Bethesda, Maryland 20014

2998 **GELLIS, SYDNEY S.; & FEINGOLD, MURRAY.** Picture of the month: Trisomy 18 (Edwards' syndrome). *American Journal of Diseases of Children*, 122(2): 149-150, 1971.

Trisomy 18, or Edwards' syndrome, is characterized by failure to thrive, marked MR and motor retardation, classical facial appearance, cardiac and skeletal abnormalities, and an extra number 18 chromosome. The classical facial appearance includes a Grecian nose, somewhat small palpebral fissures, ptosis of the eyelids, epicanthal folds, microphthalmia, low-set and malformed ears, high-arched or cleft palate, micrognathia, and prominent occiput. The disorder is thought to be due to nondisjunction which occurs during meiosis. In rare instances it may be secondary to an inherited translocation. Approximately 90% of children affected by the disorder die by the first year of life. (4 refs.) - *B. J. Grylack.*

Boston Floating Hospital
Boston, Massachusetts 02111

2999 **KARDON, NATALINE B.; BERATIS, NICHOLAS G.; HSU, LILLIAN Y. F.; MOLOSHOK, RALPH E.; & HIRSCHHORN, KURT.** 47,XXY/48,XXXY/49, XXXXY mosaicism in a 4-year-old child. *American Journal of Diseases of Children*, 122(2): 160-162, 1971.

An unusual case of triple mosaicism (XXY/XXXY/XXXXY) has been observed in a 4-year-old white boy suspected of having Klinefelter's syndrome because of a small penis and small firm testes. His developmental milestones were within normal limits, but his speech was delayed. His growth followed along the third to tenth percentile range in both height and weight. IQ at 4 years of age was normal. The mosaicism

in this case probably derived from a primary meiotic nondisjunction of X chromosomes followed by 2 additional postzygote nondisjunctions. Only 1 previous case of triple mosaicism XXY/XXXY/XXXXY has been described. In both cases, there was a predominant XXXXY cell line in peripheral leukocytes, but the patients differed clinically. (11 refs.) - *B. J. Grylack.*

Mt. Sinai School of Medicine of the
City University of New York
New York, New York 10029

3000 SHAPIRO, LAWRENCE R.; BRILL, CHARLES B.; HSU, LILLIAN Y. F.; CALVIN, MERRILL E.; & HIRSCHHORN, KURT. Deceleration of intellectual development in a XXXXY child. *American Journal of Diseases of Children*, 122(2):163-164, 1971.

A child with the XXXXY syndrome who had a normal Gesell Adaptive Maturity Level at 15 months of age was reevaluated at 26, 36, and 41 months and was found to have definite deceleration of intellectual development and consequent MR. This was the first case of a patient with the XXXXY syndrome in whom the mode of development of MR had been elucidated. Prospective observations of other patients with sex chromosome aberrations, which are known to be associated with MR, will help to confirm whether or not the MR occurs after a deceleration in intellectual development. (7 refs.) - *B. J. Grylack.*

Letchworth Village
Thiells, New York 10984

3001 SCHMICKEL, ROY D.; SILVERMAN, EUGENE M.; FLOYD, ALTON D.; PAYNE, FRANCIS E.; POOLEY, JANET M.; & BECK, MALCOLM L. A live-born infant with 69 chromosomes. *Journal of Pediatrics*, 79(1): 97-103, 1971.

A live-born infant was observed to have 69 chromosomes, and comparison of this case of triploidy with others described in the literature demonstrated a clinically recognizable syndrome which should enable the condition to be diagnosed. The available facial photographs of triploid infants were similar. The presence of colobomas, simian creases, syndactyly of third and fourth digits, high number of digital whorls, hypospadias and testicular interstitial cell hypoplasia were also common to the triploidy cases. Despite their additional 23 chromosomes, triploid cells behave remarkably like diploid cells. They do not have the increased susceptibility to an oncogenic virus which is characteristic of certain trisomic cells. Contact inhibition of triploid, diploid, and haploid cells is similar. The increased red cell antigen strength of triploid cells is heightened proportionately to the increase in gene dosage. It is felt that gene dosage must be important in embryologic development in a manner which is specific for the genes involved and by a mechanism which is not understood. (25 refs.) - *B. J. Grylack.*

University of Michigan
Ann Arbor, Michigan

3002 MICHAELS, DAVID L.; GO, SUMIO; HUMBERT, JAMES R.; DuBOIS, REUBEN S.; STEWART, JANET M.; & ELLIS, ELLIOT F. Intestinal nodular lymphoid hyperplasia, hypogammaglobulinemia, and hematologic abnormalities in a child with a ring 18 chromosome. *Journal of Pediatrics*, 79(1): 80-88, 1971.

A 12-year-old girl was seen and was reported to have intestinal nodular lymphoid hyperplasia (INLH) and severe hypogammaglobulinemia (absent IgA, low IgG and IgM) in unusual combination with a ring 18 chromosomal defect and hematologic abnormalities involving the leukocytes, erythrocytes, and platelets. Although an isolated IgA deficiency may be associated with a ring 18 chromosome, neither INLH nor hematologic abnormalities has been reported previously in association with this chromosomal defect. In this patient, the *in vitro* phagocytic capability of the neutrophils was lower than that of cells from controls. Leukocyte adhesiveness was low and was attributable directly to the neutrophils. Although the narrow hypoplasia and relative pancytopenia manifested by the patient were consistent with failure at a basic stem cell level, the lymphoid hyperplasia suggests that sufficient lymphoid precursors were generated to demonstrate the further block in normal plasma cell maturation. (29 refs.) - *B. J. Grylack.*

National Jewish Hospital
Denver, Colorado 80206

3003 **HSU, LILLIAN Y. F.; SHAPIRO, LAWRENCE R.; GERTNER, MELVIN; LIEBER, ERNEST; & HIRSCHHORN, KURT.** Trisomy 22: A clinical entity. *Journal of Pediatrics*, 79(1): 12-19, 1971.

Three female children with a characteristic facies, growth retardation, MR, microcephaly, cleft palate, micrognathia, congenital heart disease, preauricular skin tags with low-set and/or malformed ears, deformed lower extremities, appendages and sinuses, finger-like malopposed thumbs, abnormal and/or low-set nipples, and cubitus valgus were diagnosed to have trisomy 22. The peripheral leukocyte cultures of each of the 3 patients and the fibroblast culture of 1 case revealed 47 chromosomes with an extra small acrocentric chromosome, which was identical in appearance to a G-group chromosome, and normal female sex chromosome constitution (XX). Autoradiographic studies done of 2 cases with the aid of a binomial expansion tended to identify the extra chromosome as a number 22. On the basis of the phenotypic, cytogenetic, and autoradiographic findings in these 3 cases and in 10 similar patients reported in the literature, it appears that trisomy 22 is a distinct clinical entity. (26 refs.) - *B. J. Grylack.*

Mount Sinai School of Medicine
New York, New York

3004 **YANAGISAWA, S.; & HIRAOKA, K.** Familial C/G translocation in three relatives associated with severe mental retardation, short stature, unusual dermatoglyphics and other malformations. *Journal of Mental Deficiency Research*, 15(Part 2): 136-146, 1971.

Three SMR male relatives presented with short stature, unusual dermatoglyphics involving absent band/or c palmar digital triradii in all cases and arch proximal in the hallucal area in 2 of the cases, and other malformations including strabismus, prominent nasal bridge, high-arched palate, abnormal flexion of fingers, retroflexible thumbs, hypoplastic toenails, and small penis. Cytogenetically, they had marker chromosomes with a translocation between 1 of the C-group chromosomes and 1 of the G-group chromosomes resulting in partial C trisomy and partial G monosomy. These unusual anomalies were transmitted by balanced carriers. Many spontaneous abortions were recognized in the pedigree of the present Ss, and some relatives were MRs. The chromosomal aberrations with familial translocation are probably responsible for hereditary mental deficiencies. (25 refs.) - *B. J. Grylack.*

Yamaguchi University School of Medicine
Ube, Japan

3005 **MORIC-PETROVIC, SLAVKA; MARKOVIC, VJERICA; KALICANIN, P.; GARZICIC, B.; ZIVKOVIC, S.; SOLDATOVIC, B.; MARKOVIC, STEFANIJA; DESPOTOVIC, MILICA; & LACA, ZIVANA.** Cytogenetic survey of Down's syndrome in Serbia (Yugoslavia): Incidence of numerical and structural abnormalities, *Journal of Mental Deficiency Research*, 15(Part 2): 102-114, 1971.

Four hundred and thirty-six persons with Down's syndrome in Serbia, Yugoslavia, were examined clinically, and 180 of these Ss and 341 members of their families were examined cytogenetically. The results were divided into 2 groups, according to whether the mother was under 30 years of age (Group I) or over (Group II) at the birth of the mongol. Of 130 propositi of Group I, 9 translocations were found, including 3 D/G and 6 G/G, 1 of which was considered a partial G/G translocation and was discovered in a boy with Down's syndrome. Of the 3 D/G translocations, 2 occurred in sisters with Down's syndrome. No translocations were discovered in Group II. Among the 180 propositi examined, 3 cases of mosaicism were found in Group I and 1 in Group II, the features of Down's syndrome being relatively mild and intelligence being relatively high in all of them. On the basis of clinical examination and karyotypic analysis, these individuals can be considered as partial mongols. Other chromosomal anomalies in the Ss surveyed included a transmitted C/G translocation, a triple X mother, an enlarged short arm of 1 G-group chromosome in 5 families of propositi with trisomy G, a double satellite on 1 of the D-group chromosomes in the family of 1 propositus with trisomy G, satellite-like formations on 1 chromosome of group E (17 or 18), and a mongol girl with trisomy G who gave birth to a genetically normal infant. (16 refs.) - *B. J. Grylack.*

Institute for Mental Health
Belgrade, Yugoslavia

3006 **KUCERA, J.** Leukaemia and twinning tendency in families of children with Down's syndrome. *Journal of Mental Deficiency Research*, 15(Part 2): 77-80, 1971.

Children with Down's syndrome and their relatives were investigated for a possible association of Down's syndrome and leukemia within families. Twenty-one leukemic Ss were found in 801 families of children with Down's syndrome, the incidence of leukemia being more frequent in families of mothers of children with Down's syndrome than of fathers of these children (16.5 per 1,000 and 7.8 per 1,000, respectively). The evidence of twinning was significantly higher on the maternal side of the family (P=0.01). With regard to the relationship between twinning and leukemia, there is a tendency towards concordance of distribution in families. The obtained data support the hypothesis that a tendency to abnormal cell division, or to an increase in the production of chromosomal aberrations, exists within the families of mothers who have children with Down's syndrome. (6 refs.) - *B. J. Grylack.*

Institute of Care for Mother and Child
Prague 4, Czechoslovakia

3007 **GOODMAN, R. M.; KATZNELSON, M. BAT-MIRIAM; SPERO, M.; SHAKI, R.; PADEH, B.; & SADAN, N.** The question of trisomy 22 syndrome. *Journal of Pediatrics*, 79(1): 174-175, 1971. (Letter)

A 10-month-old infant admitted to a pediatric service for evaluation of multiple congenital anomalies was found to have features similar to other cases of trisomy 22 syndrome described in the literature. In addition to hypotonia, the patient suffered from mental, growth, and developmental retardation. Anomalies of the head and facies included marked asymmetry of the head, left frontal bossing, larger left ear than right, right preauricular ear tag, bilateral epicanthal folds, increased philtrum length with an accentuation of the cleft, micrognathia, high-arched palate, and hyperplasia of the gingiva. Other anomalies consisted of a short neck with mild webbing, long fingers with low-set thumbs, congenital dislocation of the right hip, right inguinal hernia, and undescended left testicle. Chromosome analysis utilizing both peripheral blood and skin showed a chromosome number of 47 with a group G trisomy. Dermatoglyphic studies of the hands showed features untypical of Down's syndrome. (8 refs.) - *B. J. Grylack.*

Tel-Hashomer Hospital
Tel-Aviv, Israel

3008 **RICHARDS, B. W.** The incidence of mongol twins. *Journal of Mental Deficiency Research*, 15(Part 2): 121-129, 1971.

The overall incidence of liveborn twinning in 1,579 sibships was ascertained through the presence of 1,596 mongol propositi. The total number of singletons and twins in the sibships, including the mongols, was 5,941. The overall incidence of liveborn twins agreed quite closely with that of the general population. Calculation of the proportions of monozygotic (MZ) and dizygotic (DZ) twins with Weinberg's method showed proportions of 0.95 MZ to 1.80 DZ in the nonmongol category and 0.46 MZ to 1.17 DZ in the mongol category. The low incidence of liveborn twins among mongols was of similar degree for both types of twin. This low incidence may be due to selection of the mongol sample from those who have survived infancy, or it may result from the difficulty that chromosomally abnormal ova could have in becoming fertilized. The analysis of the sample of mongols described did not permit any definite conclusions concerning the incidence of mongol twins at birth. (30 refs.) - *B. J. Grylack.*

St. Lawrence's Hospital
Caterham, Surrey, England

3009 **PARTINGTON, M. W.; MACDONALD, M.R.A.; & TU, J. B.** 5-Hydroxytryptophan (5-HTP) in Down's syndrome. *Developmental Medicine and Child Neurology*, 13(3): 362-372, 1971.

5-Hydroxytryptophan (5-HPT) was given in a double-blind crossover trial to 6 children with Down's syndrome and 6 other SMR children in order to assess its effect on motor behavior and neurological status. Whole blood serotonin (of which 5-HPT is a precursor) was decreased in children with Down's syndrome as had been previously noted and administration of 5-HPT did result in increased 5-hydroxy indoles in

whole blood and excretion of 5-hydroxyindole acetic acid in the urine. There was no change in the level of these products in the CSF. No marked or consistent changes in motor behavior or neurological status were noted. 5-HPT cannot be considered a specific treatment for Down's syndrome. (14 refs.) - *E. L. Rowan.*

Queen's University Department of Paediatrics
Ontario, Canada

3010 **BARNET, ANN B.; OHLRICH, ELIZABETH S.; & SHANKS, BETTY L.** EEG evoked responses to repetitive auditory stimulation in normal and Down's syndrome infants. *Developmental Medicine and Child Neurology*, 13(3): 321-329, 1971.

Auditory evoked responses were quantitatively different in normal infants and infants with Down's syndrome (Trisomy 21 on karyotype). Groups of 1, 6, and 12-month-olds were tested. Normal infants aged 6 and 12 months showed progressive response decrement on repetitive stimulation. Normal neonates and Down's syndrome infants in all groups failed to show this decrement. A prominent long latency positive component (P3 wave) was also noted in older normals but not in neonates or infants with Down's syndrome. The mental defect in Down's syndrome is related to an inability to adapt and moderate response to repetitive stimulation. (33 refs.) - *E. L. Rowan.*

Children's Hospital
Washington, D. C. 20009

3011 **McCOY, E. E.; & NANCE, JOANNE L.** Lipid composition and synthesis of isolated polymorphonuclear leucocytes in Down's syndrome. *Journal of Mental Deficiency Research*, 15(1): 1-11, 1971.

Comparison of individuals with Down's syndrome and a control group indicated that Ss with Down's syndrome had a decreased amount of total lipid which was primarily in the cholesterol and in several phospholipid fractions. Ss were 20 males (CA range 15-25 yrs) with Down's syndrome and 14 male MRs who did not present Down's syndrome. Tissue source for the study of lipid composition and synthesis was isolated polymorphonuclear leukocytes. Since phospholipids have an important function in relation to cell structure and function, the changes observed in this study may be used to explain some of the biochemical differences noted in the leukocytes of individuals with Down's syndrome. (25 refs.) - *J. K. Wyatt.*

University of Alberta School of Medicine
Edmonton, Alberta, Canada

3012 **ZDZIENICKA, E.; & KACPERSKA, E.** Occurrence of Australia antigen in patients with Down's syndrome in Poland. *Journal of Mental Deficiency Research*, 15(1): 30-37, 1971.

Investigation of the occurrence of Australia antigen in patients with Down's syndrome in Poland revealed frequencies of 7% in Down's syndrome Ss and 2% in control group Ss. There were 98 Ss with Down's syndrome (52 males, 46 females; age range 8 to 27 yrs) and 98 Ss who evidenced other types of MR and were matched for sex, age, and environment. Institutionalized Down's syndrome Ss had a frequency of 8.3%; the frequency for outpatients was 5.2%. Between-group differences were not significant. Frequency of Australia antigen was not related to size of institution. The antigen was detected in 2 of 52 males and 4 of 46 females with Down's syndrome. Highest frequency was in the 10- to 19-year-old group. Presence of the Australia antigen was significantly associated with elevated SGPT level in Down's syndrome Ss. Frequency of Australia antigen in Down's syndrome Ss was significantly ($p<.001$) greater in Down's syndrome Ss than in the normal population. (21 refs.) - *J. K. Wyatt.*

Pruszkow Psychiatric Hospital
Pruszkow, Poland

3013 **RICHARDS, B. W.; RUNDLE, A. T.; HATTON, W. M.; & STEWART, A.** G-Group ring chromosome in a mentally subnormal girl. *Journal of Mental Deficiency Research*, 15(1): 61-72, 1971.

Many characteristics exhibited by a PMR girl with a G-group ring chromosome were opposite

to those of mongolism. She had increased muscle tone, a restless and excitable temperament, and increased blood level creatinine phosphokinase. Levels of serum uric acid and galactose-1-uridyl transferase were normal. Her cephalic index was similar to that found in Down's syndrome. Comparison of the 3 cases of G-group ring chromosome with stable rings and no mosaicism that have been reported does not provide a characteristic clinical syndrome which could be used to diagnose the presence of a ring-G-chromosome with terminal deletions. (31 refs.) - *J. K. Wyatt.*

St. Lawrence's Hospital
Caterham, Surrey, England

3014 **SASAKI, MOTOMICHI; IKEUCHI, TATSURO; OBARA, YOSHITAKA; HAYATA, ISAMU; MORI, MICHIKO; & KOHNO, SEI-ICHI.** Chromosome studies in early embryogenesis. *American Journal of Obstetrics and Gynecology*, 3(1): 8-12, 1971.

To provide basic information for cytogenetic studies, congenital anomalies, and spontaneous abortion, the frequency of chromosome abnormalities in early development was surveyed in Sapporo, Japan, in 829 randomly collected curetted conceptuses of 4-23 weeks. Chromosome and sex-chromatin studies were made on cultured embryonic and/or extraembryonic tissue and anamnestic data were obtained by interview. Three cases of X monosomy (45,X), 1 of X trisomy (47, XXX), 3 of sex chromosome mosaicism (45,X/46,XY; 45X/46XX; 46,XY/47,XXY), 3 of autosomal trisomies (47,XX, D+; 47,XX, E+, 47XY,C), and 1 case of translocation (45,XY,D-,t(DqDq)+, 12 males with an unusually long Y chromosome, 6 cases with an enlarged short arm in one of acrocentrics, and 1 each case with a heteromorphic pair of either No. 1 or 16 were found. A high incidence of chromosome breakage occurred in 9 cases, which may relate it to fetal risk. Tetraploidy was observed in 6 cases. Sex ratio showed an excess of females but there was gradual reversal with decreasing embryonic age. Incidence of aneuploid individuals was 1.2% for the average gestational age of 10 weeks, a value twice as high as the incidence at birth. Failure of chromosome analysis in 37% of the specimens suggests that the abnormalities observed might be minimal. (16 refs.) - *R. Seevaratnam.*

Hokkaido University
Sapporo, Japan

3015 **MOTL, MARY LOU; & OPITZ, J. M.** Studies of malformation syndromes XXVA: Phenotypic and genetic studies of the Brachmann-de Lange syndrome. *Human Heredity*, 21(1): 1-16, 1971.

Analysis of 180 cases of the Brachmann-De Lange syndrome in terms of significant phenotypic variability and genealogical and cytogenetic data pertinent to etiology revealed that a significantly greater number of anomalies is not correlated with increased severity of MR in cases reported by physicians, but that the difference between the proportion of abnormal to normal traits reported is significant. Characteristics of the syndrome include growth failure, MR, small head, small nose, downward curving upper lip, low set ears, low hairline, long thick curly eyelashes, upper limb deformities, dermatoglyphic anomalies and simian creases, genital anomalies, low-pitched voice, occasional cleft palate, heart murmurs, and eye defects. Its complete phenotype, natural history, and incidence are unknown, and a specific biochemical defect has not been discovered. The study revealed no etiological effects of birth order, parity, parental age, unusual fertility, increased incidence of gestational abnormalities, or parental consanguinity. Autosomal recessive inheritance with partial homozygote lethality is suggested as a possible etiology, the lethal effect arising from chromosome breakage shortly after fertilization or during early cleavage divisions and producing structural changes or chromosome losses in the homozygotes. (71 refs.) - *N. Jarvis.*

University of Wisconsin
Madison, Wisconsin

3016 **MEHES, K.; & SULYOK, E.** Sex chromatin aberrations among children with severe mental retardation. *Human Heredity*, 21(1): 54-56, 1971.

A sex chromatin survey of 1,000 inst MR children in Pecs, Hungary, revealed a high number of aberrations among SMR, a population of 406 accounting for 9 of the 14 aberrations observed. This finding is in contrast to the assertion that the number of sex chromatin aberrations found

among the grossly defective is not significantly different from the number found in the normal population. Although the number of patients examined is relatively small, findings suggest that the frequency of sex chromatin aberration among SMR is greater than commonly believed. (7 refs.) - *N. Jarvis.*

University Medical School
Pecs, Hungary

3017 **SACHDEVA, SHANTA; WODNICKI, JANICE; & SMITH, J. F.** Fluorescent chromosomes of a tandem translocation in a mongol patient. *Journal of Mental Deficiency Research*, 15(3): 181-184, 1971.

A report is made of a female mongol patient (CA 2 months) having a tandem translocation of 2 number 21 chromosomes. Chromosomes were examined using a Leitz fluorescent microscope. S had 46 chromosomes, all of which appeared normal except for group G. She had only 3 group G chromosomes and a large acrocentric chromosome slightly smaller than the D group chromosomes. The fluorescent patterns for G group chromosomes showed 1 G chromosome with a fluorescing band in the long arm. The other 2 G group chromosomes (assumed to be number 22s) showed very weak fluorescence of the long arms. The large acrocentric chromosome showed 2 prominent bands fluorescing in its long arms. The fluorescing patterns of the 6 D group chromosomes showed 3 pairs of identical fluorescing patterns all different from the fluorescing of the large acrocentric chromosome. The identification of satellites on the short and long arms of the tandem chromosome would suggest that it was formed by the fusion of the long arms of 2 G chromosomes. The double prominent fluorescing bands on the tandem chromosome suggest that the long arms of 2 number 21 chromosomes have fused. It is much too early yet to be certain that the fluorescing patterns of numbers 21 and 22 chromosomes will be similar in all mongols. (12 refs.) - *T. C. Turner.*

Loyola University of Chicago
Stritch School of Medicine
Maywood, Illinois 60153

3018 **FIELDS, DONALD L.; & GIBSON, DAVID.** Forecasting mental growth for at-home mongols (Down's syndrome). *Journal of Mental Deficiency Research*, 15(3): 163-168, 1971.

A study is reported, the specific aims of which were to: (a) test the potency of an earlier (Gibson, 1966) mental growth forecast instrument for community-based mongols; (b) determine if additional variables increase the relationship between developmental markers and maturity MA over the earlier 3-variable formula; (c) explore the possibility that a factor analytic solution for developmental markers in mongolism might apportion the differential effects of physiological and environmental influence on intellectual maturity levels for the syndrome. Ss were 22 adult mongols. Psychometrics were based on the Stanford-Binet intelligence test. Multiple regression and principal components analyses on the 5 predictors were produced by computer. Gibson's weightings for age in months at teething, walking and talking were employed to estimate MA for all Ss. A principal components analysis of the unchanged correlation matrix was then conducted and the 2 most significant factors were rotated to Varimax criteria. Three Tables present Gibson's variables with MA, the power of his markers and expanded formula for MA and factor loadings for the rotated factor matrix. The findings indicate that prediction for a community based mongol is as reliable as for an institutional mongol. The factor matrix outcome indicates tentatively that early growth in the mongol is environmentally susceptible. The establishment of a reliable forecast instrument based on early growth indicators would aid in designing suitable programs to prepare the mongol for self-sufficient adulthood with maximum capacity. (17 refs.) - *T. C. Turner.*

York University
Ontario, Canada

3019 **DALLAPICCOLA, B.** Australia antigen and Down's syndrome. *Lancet*, 2(7723): 548, 1971. (Letter)

Studies of 73 Ss (49 males, 24 females) with trisomy-21 Down's syndrome, 1 male with a G/G translocation, and 1 mosaic male revealed no Australia antigen in the sera of the latter two. Data confirmed a decreased incidence of Au in females. No conclusion could be drawn between association of Au and Down's syndrome. (2 refs.) - *B. Berman.*

Clinica Medica
Ferrara, Italy

3020 **AHMAD, K. N.; DYKES, J. R. W.; FERGUSON-SMITH, M. A.; *LENNOX, B.; & MACK, W. S.** Leydig cell volume in chromatin-positive Klinefelter's syndrome. *Journal of Clinical Endocrinology and Metabolism*, 33(3): 517-520, 1971.

Histometric measurements of Leydig cell volume (Vld) in 50 cases of chromatin-positive Klinefelter's syndrome (26 were plain XXY) revealed no cell hyperplasia in this condition (mean Vld of 0.82 ml was within normal limits). Three cases with very low Vld had complete hyalinization of all or nearly all tubules. The single instance of very high Vld apparently was caused by earlier removal of the other testis. There is evidence that the pituitary control of tubules and Leydig cells reflects two totally different processes. At least some of the changes in necropsy XXY testes result from testosterone treatment. (7 refs.) - *B. Berman.*

*Western Infirmary
Glasgow W1, Scotland

3021 **RIDLER, M.A.C.; SCHWINGER, B.; RITTNER, CH.; & CROSSEN, P. E.** Banding patterns of metaphase chromosomes in Down's syndrome. *Lancet*, 2(7724): 603, 1971. (Letters)

Ridler rejects Baikie's complaint about the classification of the extra chromosome in Down's syndrome and states that when that chromosome is identified by the Giemsa banding method it is indistinguishable from the smaller pair of G-group chromosomes. Schwinger and Rittner claim that, in 41 Down's syndrome Ss investigated with fluorescence microscopy, the extra chromosome was no. 21, not no. 22, as stated by Ridler. Crossen, also, using a technique similar to Giemsa binding, identifies the chromosome as no. 21. (9 refs.) - *B. Berman.*

Harperbury Hospital
St. Albans, Hertfordshire, England

3022 **HAMERTON, JOHN L.** Banding patterns of metaphase chromosomes in Down's syndrome. *Lancet*, 2(7726): 709, 1971. (Letter)

It has been resolved by international agreement at the Fourth International Conference on Standardization in Human Cytogenetics (held in Paris September 2 to 4, 1971) to define chromosome number 21 by its fluorescence pattern as the chromosome that is associated with Down's syndrome and is smaller than number 22. By definition, therefore, the ph chromosome becomes number 22. It is hoped that this agreement will be accepted by the majority of cytogeneticists. (1 ref.) - *B. J. Grylack.*

Children's Hospital
Winnipeg, Canada

3023 **MILUNSKY, AUBREY; ATKINS, LEONARD; & LITTLEFIELD, JOHN W.** Polyploidy in prenatal genetic diagnosis. *Journal of Pediatrics*, 79(2): 303-305, 1971.

Occurrence of polyploidy (increase in the number of chromosomes per cell in multiples of the basic number) was studied in 24 pregnancies by obtaining amniotic fluid samples for prenatal genetic diagnosis between 16 and 20 weeks of gestation. In 16 of the 24 consecutive cases, polyploidy (mostly tetraploid but also some triploid and hexaploid cells) was found in the cultured amniotic fluid cells. Frequency of such cells was 4-83.3%. Nonetheless, 14 clinically normal infants have been delivered, including 9 pregnancies with 4-43% polyploidy. Extensive polyploidy in amniotic fluid cell cultures thus appears to be quite compatible with the birth of clinically normal children. Since prenatal diagnosis of chromosomal anomalies by means of cultured amniotic fluid cells is becoming more widely available, one should be wary of mistaken interpretations of the significance of polyploidy in these cells in order not to cause undue interruption of a pregnancy involving a normal fetus. (9 refs.) - *K. L. Woodworth.*

200 Trapelo Road
Waltham, Massachusetts 02154

3024 **COLEMAN, MARY.** Infantile spasms associated with 5-hydroxytryptophan administration in patients with Down's syndrome. *Neurology*, 21(9): 911-919, 1971.

Oral administration of 5-hydroxytryptophan (5-HTP) to patients with Down's syndrome resulted in improved muscular tone and activity

level and suppression of buccolingual dyskinesias, but in some it appeared to induce a seizure disorder resembling the infantile spasm syndrome. The factors contributing to this onset are discussed and the subject of serotonin and seizures is reviewed: a rapid rate of 5-HTP increase in a short time period is conducive to the induction of seizures; the L form of 5-HTP is more likely to induce the syndrome than the D,L form; the D,L form is associated with seizure activity usually in cases where 5-HTP treatment is initiated after the neonatal period; in most cases pyridoxine does not seem to prevent or reverse the syndrome; once the EEG has become abnormal, the prognosis for stopping the seizures by reducing/stopping 5-HTP is poor; review of clinical histories showed several patients to be sensitive to 5-HTP; no evidence of a familial disposing factor was found in any patient. The rarity of infantile spasm plus the high frequency - 15 percent (9 of 60) - of seizures in patients in the study and cessation or amelioration of symptoms by decreasing 5-HTP is strong evidence that chronic administration of 5-HTP was an etiological factor in the seizure activity reported. The possible relevance of 5-HT (serotonin) for seizure disorders in general needs further study. (34 refs.) - *T. C. Turner.*

Children's Hospital
Washington, D. C. 20009

3025 **BODENSTEINER, J. B.; & ZELLWEGER, H.** Mongolism preventable by amniocentesis. *Clinical Pediatrics,* 10(10): 554-556, 1971.

Prenatal cytogenetics is discussed with primary reference to mongolism and the number of high risk pregnancies within an American population of 3 million people (population of the state of Iowa) with approximately 50,000 live births per year. Five groups of high risk women are: women 40 and over; 35-40 with a previous mongoloid child; mongoloid women; parental mosaicism with a 21 trisomic cell population; familial translocation (G/G & D/G). Two percent of children born to Group 1 mothers are mongoloid; 20 cases could be detected by amniocentesis of each 1000 Group 1 women. Group 2 women have a 3 times higher risk than same-age women without a previous mongoloid child. The number of high risk Group 3 women is negligible, since there is a great amount of prenatal wastage. The risk of Group 4 women having a mongoloid child depends on the prevalence of 21 trisomic spermatogonia or ovogonia. In Group 5 the risk is 100 percent if 1 parent carries a balanced 21/21 translocation. A program to identify cases of mongolism would cost about $200,000 per year. The lifetime expense of each mongoloid patient in a State institution is about $180,000, so prevention of mongoloids would lead to savings 10 times greater than the cost of the entire diagnostic operation. (8 refs.) - *T. C. Turner.*

University of Iowa Department of Pediatrics
Iowa City, Iowa 52240

3026 **MATTHIES, FRED; MACDIARMID, W. D.; RALLISON, MARVIN L.; & TYLER, FRANK H.** Renal anomalies in Turner's syndrome. *Clinical Pediatrics,* 10(10): 561-565, 1971.

Renal abnormalities in patients with Turner's syndrome are discussed. Ss were 15 children and young adults with the syndrome. Each S had buccal sex chromatin studies and cultures of circulating lymphocytes for chromosome analysis; a history of urinary complaint and pyelogram were also taken. Every S had evidence of at least 1 line of cells compatible with a chromosome complement of 45,X. Eleven Ss had structural or positional abnormalities of kidney and collecting system, or both; 3 had a horseshoe kidney. Other structural abnormalities were: ureteropelvic obstruction, double collecting system, bifid renal pelvis. Anomaly of renal position was assessed with respect to malrotation or to level of ascent in relation to the vertebrae. No patients had impaired renal function. Two of 4 with a history of urinary complaint had impaired pelvic or ureteral drainage. The lack of clinical importance of the renal abnormalities in the Ss is commented upon; none had evident impaired renal function. Further observations of the Ss were warranted, but no treatment was deemed necessary. However, 8 of 12 Ss without horseshoe kidney had renal positional anomalies similar to it (low placement and malrotation) so, in Turner's syndrome, horseshoe kidney may represent the fullest expression of a continuum of morphologic abnormalities, caused by varying degrees of failure of the "positive chemotaxis" mechanism during migration of the kidney in embryogenesis. (25 refs.) - *T. C. Turner.*

Stanford University School of Medicine
Stanford, California 94305

3027 **SYLVESTER, P. E.; RICHARDS, B. W.; RUNDLE, A. T.; & STEWART, A.** Pathological observations on a male patient with D-ring chromosome. *Journal of Mental Deficiency Research*, 15(3): 207-223, 1971.

The nineteenth case of D-ring chromosome (the first in which results of a post-mortem investigation are available) is reported. S was an epileptic, MR male (CA 18 years, MA 19 months) who died of bronchopneumonia. He was short, obese, sexually underdeveloped, with small head; heart, lungs, sense organs and cranial nerves were normal. S had 46 chromosomes and in 5 percent of cells examined there was a double sized dicentric ring, considered a D-ring. Necropsy showed good body nutrition, congested lungs, normal heart, stomach, intestines, greatly enlarged liver and spleen. Brain stem and cerebellum were well-formed, with mild dilation of the lateral ventricles; nerve cells had normal shape, gliosis was absent and there were no senile plaques. Hypoplasia was present in the middle lobe of the cerebellum. In the thoracic region of the spinal cord on the right side, the cells of the nucleus dorsalis were deficient. Findings on this S are compared with other examples of Ss with D-ring chromosome: All Ss had severe mental subnormality, retardation of physical growth, prominent nose bridge and large ears; epilepsy and ataxia are often present. (32 refs.) - *T. C. Turner.*

St. Lawrence's Hospital
Caterham, Surrey, England

3028 **FIALKOW, P. J.; BLUMBERG, B. S.; LONDON, W. T.; SUTNICK, A. I.; & THULINE, H. C.** Thyroid antibodies and Australia antigen in Down's Syndrome. *Journal of Mental Deficiency Research*, 15(3): 177-180, 1971.

Australia antigen and thyroid antibodies in 106 institutionalized and 14 at-home Ss with Down's Syndrome (DS) were studied to test the hypothesis that the presence of thyroid antibodies in Ss with DS is associated with Australia antigen. Sera were tested for thyroid antibodies with specificity for thyroglobulin and for thyroid cytoplasmic antigen. Thyroid antibodies were detected in the sera of 36 institutionalized Ss; Australia antigen was found in 4 of those 36 and in 16 of the 70 sera from Ss with negative thyroid antibody tests. A statistically significant association between the presence of Australia antigen and thyroid antibodies in Ss with DS was not found. The data therefore do not support the hypothesis that thyroid antibodies in patients with DS arise as a consequence of active hepatitis associated with Australia antigen. (14 refs.) - *T. C. Turner.*

University of Washington Department of Medicine
Seattle, Washington 98195

3029 **MACINTYRE, M. NEIL; WALDEN, DAVID B.; & HEMPEL, JOANNE M.** Tertiary trisomy in a human kindred containing an E/G translocation. *American Journal of Human Genetics*, 23(5): 431-441, 1971.

An abnormal karyotype (a normal complement plus a satellited, minute chromosome) was found in a congenitally malformed proband whose mother carried a balanced E/G translocation (this turned out to be a 17/G) with the marker G equivalent to the proband's minute chromosome. Breaking and rejoining of the interchange segments presumably occurred in a greatgrandparent germ line or in an early somatic cell. The translocation appeared in a sibling and in the maternal grandmother. Proband's karyotype apparently came from nondisjunction and did not represent an expected segregation from a balanced translocation. (14 refs.) - *B. Berman.*

Case Western Reserve University
Cleveland, Ohio

3030 **GENDEL, EDWARD.** Turner's syndrome karyotypes. *Journal of the American Medical Association*, 215(7): 1160, 1971. (Letter)

A letter describing a woman with some of the features of Turner's syndrome is illustrated with a karyotype in which both the third pair of chromosomes in Group A and the sex chromosome pair are mislabeled. Barr body or sex chromatin studies ought to include the size of the sex chromatin body, since a smaller than normal one can produce a syndrome like Turner's syndrome, but with functioning ovaries. (1 ref.) - *S. Glinsky.*

No address

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3031 **KRMPOTIC, EVA; & HARDIN, MARY BETH.** Secondary nondisjunction causing regular trisomy 21 in the offspring of a mosaic trisomy 21 mother. *American Journal of Obstetrics and Gynecology*, 110(4): 589-590, 1971.

A case of Down's syndrome was transmitted through maternal mosaicism, which was detected by cytogenetic diagnosis. The propositus was a baby girl who was diagnosed to have regular trisomy 21. Chromosome studies of the mother's blood revealed a mosaic trisomy 21, although she exhibited no clinical signs of Down's syndrome or MR. The origin of the syndrome in the mother can be attributed to secondary nondisjunction during meiosis. Careful cytogenetic examination is important due to the high ratio of affected offspring (1:1) from parents with mosaic trisomy 21. (2 refs.) - *S. Glinsky.*

Mount Sinai Hospital Medical Center
Chicago, Illinois

MEDICAL ASPECTS – Miscellany

3032 **DUMARS, KENNETH W., JR.** Parental drug usage: Effect upon chromosomes of progeny. *Pediatrics*, 47(6): 1037-1041, 1971.

Comparison of clinical and chromosomal karyotype examinations of infants born to illicit users of LSD and a control population did not provide evidence of increased chromosomal breakage or structural rearrangement among Ss of the experimental group. Interviews prior to delivery provided information on timing and extent of drug usage of both parents of 47 infants from a population of over 1,000 consecutive infants relinquished for adoption. Data on the extent of usage covered as well as possible whether the drug was used before or during pregnancy or both, the extent of use of drugs in addition to LSD, and the frequency of use by both parents. Approximation of dose levels was not possible. Of the 47 test infants, 8 had clinical abnormalities, but of 41 of the test Ss and 20 control Ss examined karyotypically, no statistical differences in chromosome abnormalities between the 2 groups appeared. All mothers and putative fathers of the test Ss used a multiplicity of drugs in addition to LSD. Results indicate that a history of parental drug usage does not constitute a valid reason for failure of an agency or a family to proceed with adoption. (30 refs.) - *M. S. Fish.*

University of California,
College of Medicine
Irvine, California 92664

3033 Teratologists united. *Nature*, 231(5298): 86, 1971.

The recently formed European Teratology Society met recently to discuss various facets of this field. The presence at this meeting of clinicians, epidemiologists, and experimental scientists promises to aid in future improvement of the quality of research in this area. Symposium participants stated the issues related to the etiology of birth defects but were able to provide little new information in this field. Only a few genetic or environmental factors have so far been related to the cause of human congenital malformations. Some experiments have implicated nutritional factors, and the incidence of CNS defects has been related to softness of drinking water. The symposium also examined the problem of screening drugs for teratogenic effects. The participants concluded that current screening methods reduce the potential hazards of drugs such as thalidomide; however, present regulations may be overcautious in that they may ban potentially useful drugs. Species variability came up for consideration, but the participants did not reach a conclusion on the need to utilize primates in this research, although they felt that possible teratogens should be tested in some nonrodent species. (No refs.) - *M. S. Fish.*

3034 **HSIA, Y. E.; BRATU, M.; & HERBORDT, A.** Genetics of the Meckel syndrome (dysencephalia splanchnocystica). *Pediatrics*, 48(2): 237-247, 1971.

Seven cases of the Meckel syndrome were discovered in 2 families, and the genetic basis of this syndrome was confirmed. The diagnostic criteria for the Meckel syndrome require at least 2 of the triad of occipital encephalocele, polydactyly, and cystic kidneys, with other major features of the disorder such as cleft palate and microcephaly. There was much variability in the features of the syndrome from 1 case to another, but the diagnosis was clear in most cases because of the unique combination of characteristics comprising the syndrome. The recurrence of the Meckel syndrome within sibships, affecting both sexes, its occurrence in consanguineous matings, and its concurrence in monozygous twins have provided convincing evidence that it is a recessively inherited condition determined by a single mutant autosomal gene, which is expressed in the homozygote. The basic lesion in the Meckel syndrome may affect an embryonic organizer. (24 refs.) - *B. J. Grylack.*

Yale University School of Medicine
New Haven, Connecticut 06510

3035 **KRAVITZ, HARVEY; & BOEHM, JOHN J.** Rhythmic habit patterns in infancy: Their sequence, age of onset and frequency. *Child Development,* 42(2): 399-412, 1971.

A systematic study was made of rhythmic habit patterns of infants. Handsucking was studied in 140 newborn normal Ss and in 79 newborn Ss with low Apgar scores and neonatal disease. Two hundred older Ss were studied for other rhythmic habit patterns. Handsucking was observed much earlier in normal than abnormal groups. Onset of rhythmic habit patterns was markedly delayed in retardates. These patterns may be valuable in the early diagnosis of developmental retardation; they may also be useful in illuminating maturation processes and development of motor skills and learning in infants. (38 refs.) - *T. C. Turner.*

Children's Memorial Hospital
Chicago, Illinois 60614

3036 **GILMOUR, DOUGLAS G.; BLOOM, ARTHUR D.; LELE, KUSUM P.; ROBBINS, EDWIN S.; & MAXIMILIAN, CONSTANTINE.** Chromosomal aberrations in users of psychoactive drugs. *Archives of General Psychiatry,* 24(3): 268-272, 1971.

Chromosome studies were carried out on 27 male and 29 female drug-users and 7 male and 9 female nondrug-users in order to determine the presence of structural abnormalities. With the exception of controls and those who used marihuana lightly, all drug users were found to have an increased incidence of chromosomal aberrations. The increases were not general in any class of drug users but were accounted for almost entirely by a few individuals in each group with 2 or more aberrant peripheral blood leukocytes out of 50 examined. All the structural chromosomal aberrations described here and in other experiments have been sporadic in occurrence. The occasional aberrations seen here were noted only in somatic cells, and no inferences can be made concerning the basis of their presence, meiotic chromosome effects, or transmission to progeny. (12 refs.) - *B. J. Grylack.*

New York University School of Medicine
New York, New York 10016

3037 **BELL, W. E.; & McCORMICK, W. F.** Striatopallidonigral degeneration. Report of a case in a 15-year-old girl. *Archives of Disease in Childhood,* 46(248):533-538, 1971.

A 15-year-old female who presented with a sporadic, progressive illness manifested by unilateral limb rigidity, dystonia, dysarthria, and a decline in intellectual capacity was given a diagnosis of striatopallidonigral degeneration after specific neuropathological findings were recorded at autopsy. The globi pallidi were shrunken and revealed a pronounced yellow discoloration. Severe neuronal loss was present in the caudata and putamena bilaterally, and moderate depletion of neurons was noted in the globi pallidi. There was intense astrogliosis in the caudate nuclei, putamena, and moderate gliosis in the globi pallidi. Neuronal loss was also present in the reticular zone of the substantia nigra with a moderate degree of replacement gliosis. While this case shared certain pathological characteristics with striatonigral degeneration, the differences seemed sufficient to consider it as a separate syndrome. (9 refs.) - *B. J. Grylack.*

University Hospital
Iowa City, Iowa 52240

3038 **WORSTER-DROUGHT, C.** An unusual form of acquired aphasia in children. *Developmental Medicine and Child Neurology*, 13(5): 563-571, 1971.

An unusual disorder was observed in 14 children, acquired receptive aphasia appearing as the outstanding symptom. After perfectly normal development, the onset of the disorder occurred in most cases between 4 and 6 years of age, usually with 1 or a few epileptiform attacks, which did not persist even in the absence of anticonvulsant treatment. A degree of deafness was evident in some cases and was followed, in sequence, by a gradual loss of comprehension of spoken language and a failure of executive speech, although any deafness gradually improved. Some hearing loss persisted in 5 cases but was relatively mild at the level of no more than 20 to 30 decibels' loss. Intelligence was not impaired. Apart from the aphasia and the occasional slight hearing loss, the children did not manifest any abnormal neurological signs. Common features also included normal copying of written material but rather primitive drawing, normal picture-arrangement for story sequences, and rapidity in associating printed and written material. The disorder may be a form of low-grade selective encephalitis, possibly of the autoimmune variety. (2 refs.) - *B. J. Grylack.*

Moor House School
Oxted, Surrey, England

DEVELOPMENTAL ASPECTS – Physical

3039 **AUXTER, DANIEL.** Motor skill development in the profoundly retarded. *American Institute for Mental Studies Training School Bulletin*, 68(1): 5-9, 1971.

Several procedures to provide greater motor function for the PMR who are in a nonambulatory state are described. Ss were 12 PMR persons (CA 12-30 years, MA 6-18 months). Programs were designed to increase the range of motion of contracted joints, develop strength of antigravity muscles directly related to upright locomotion, provide considerable movement so that proprioceptive stimulation would make the patients aware of the movement capabilities of their bodies, and attempt to integrate the movement of various joints of the body so that purposive skills development would eventually be achieved. The program's results indicate that motor function can be improved in the PMR. (1 ref.) - *T. C. Turner.*

Slippery Rock State College
Slippery Rock, Pennsylvania

3040 **GENDREAU, PAUL; & SUBOSKI, MILTON D.** Intelligence and age in discrimination conditioning of the eyelid response. *Journal of Experimental Psychology*, 89(2): 379-382, 1971.

Discrimination-eyelid conditioning tests on 65 penitentiary inmates (divided into 2 IQ groups: (a)mean IQ 88.7; (b)mean IQ 119.8, and 2 age groups:Low-Age, 21.2 years; High-Age, 36.8 years) showed better discrimination among the High IQs and more responsiveness among the Low-Age group. There was no difference in discrimination in relation to age. Better discrimination in the High-IQ group was a result of lower response levels to CS-, considerably higher CR/UCR response slope ratios, and a higher degree of classified voluntary-form responses. (11 refs.) - *B. Berman.*

Trent University
Peterborough, Ontario, Canada

3041 **JAMMES, J. L.; & *ROSENBERGER, P. B.** Rocking behavior and heart rate in the mentally retarded. *Journal of Nervous and Mental Disease*, 153(1): 57-59, 1971.

Rocking behavior and heart rate were studied in 101 MR patients from various diagnostic groups by administering cardiotropic drugs in an attempt to determine what controls the rate of rocking behavior. Epinephrine hydrochloride and ouabain were the drugs employed. Patients were examined before and after drug application. An excellent correlation between heart rate and rocking frequency was observed in all cases. In 80 patients, the ratio of heart rate to rocking frequency was in the range of 1:1. In 16 patients, this ratio was of the order of 2:1. In all these cases, the response of heart rate to the drugs was in the predicted direction, and rocking rate showed a corresponding change. A common underlying autonomic control of heart and rocking rate and the ballistic effect of the cardiac pulse pressure are 2 theories of the observed interrelationship. (8 refs.) - *B. J. Grylack.*

*Walter E. Fernald State School
Waverly, Massachusetts

3042 **CUMMING, G. R.; GOULDING, D.; & BAGGLEY, G.** Working capacity of deaf, and visually and mentally handicapped children. *Archives of Disease in Childhood*, 46(248): 490-494, 1971.

The physical working capacity and maximum oxygen uptake were determined in 61 deaf, 22 visually handicapped, and 67 MR children ranging in age from 8 to 17 years. With the use of bicycle ergometers, two thirds of the deaf males were found to be above the fifteenth percentile. Of the retarded males, almost half were above the fiftieth percentile, and one quarter were below the fifteenth percentile. Of the visually handicapped males, none was above the fiftieth percentile, and more than one third were below the fifteenth percentile. The distribution for the retarded girls was the same as for retarded boys. Half of the visually handicapped girls were above the fiftieth percentile. The mean maximum oxygen uptake was similar among the deaf and visually handicapped boys, while deaf girls had higher values than visually handicapped girls. The differences in the fitness of the 3 groups studied may be related to differences in habitual physical activity which began at an early age. The deaf had higher capacities than the normal population, MRs demonstrated the same capacities, and the visually handicapped had lower capacities. (7 refs.) - *B. J. Grylack.*

Children's Hospital of Winnipeg
Winnipeg 3, Manitoba, Canada

3043 **CROSS, K. W.; HEY, E. N.; KENNAIRD, D. L.; LEWIS, SHEILA R.; & URICH, H.** Lack of temperature control in infants with abnormalities of central nervous system. *Archives of Disease in Childhood,* 46(248): 437-443, 1971.

Observations were made on temperature control in 11 infants with abnormalities of their central nervous systems by means of testing the effect of an adequate stimulus, heat or cold, on the effector organs. Four infants, described as normothermic, sweated normally to thermal stimuli at the time of investigation, and 3 infants who were studied showed the normal changes in thermal circulation index in response to variation in environmental temperature. All of these infants had a hypothalamus with normal pathways present at autopsy. The other 7 infants were judged to be poikilothermic, and autopsy revealed an absent hypothalamus or a hypothalamus present but with defective or absent connections with lateral horn cells. The evidence was consistent with the view that lack of temperature control was associated with developmental defects in the region of the hypothalamus or its connections. (28 refs.) - *B. J. Grylack.*

London Hospital Medical College
London E1 2AD, England

3044 **FOSTER, K. G.; HEY, E. N.; & O'CONNELL, BRIDGET.** Sweat function in babies with defects of the central nervous system. *Archives of Disease in Childhood,* 46(248): 444-451, 1971.

Sweat function was studied in 50 full term infants with central nervous system malformation in order to assess the possibility that sweat glands might only develop the ability to respond to direct chemical stimulation if functionally innervated *in utero.* Tests showed that babies with lumbar meningomyelocele responded to acetylcholine in the same way as healthy term babies above the level of the spinal lesion. In infants with gross disorganization of the basal ganglia or brainstem, as observed at autopsy, and in babies with anencephaly, there was no sweat response anywhere to thermal or chemical stimulation. The absence of a sweat response to direct chemical stimulation in a term infant at birth suggests the presence of a congenital defect involving the sympathetic nerves or their associated tracts in the spinal cord or brain stem. (40 refs.) - *B. J. Grylack.*

London School of Hygiene and Tropical Medicine
London W. C. 1, England

3045 **FAGLIONI, PIETRO; SCOTTI, GIUSEPPE; & SPINNLER, HANS.** The performance of brain-damaged patients in spatial localization of visual and tactile stimuli. *Brain,* 94(3): 443-454, 1971.

The impairment produced by brain lesions on the ability to correctly localize points in extrapersonal space is investigated. Two experiments were carried out and 2 sets of data gathered: 1) Ss were 265 patients with unilateral damage and 2) 121 patients with Left or Right hemisphere damage who were to reproduce the spatial arrangement of simple patterns (Visual and/or Tactile Crosses Tests) and whose performance was related to the hemisphere side of the lesion and to the presence or absence of retrochiasmatic

visual field defects. The results of the first experiment point to a differential effect of the locus of the lesion on visually-guided spatial localization, according to the half field of gaze taken into consideration. In the homolateral field the presence of visual field defect has the same detrimental effect; in the contralateral half-field the impairment produced by lesions involving the central optic pathways in the Right hemisphere is much greater than that produced by corresponding lesions of the Left hemisphere, reflecting the asymmetry of the 2 halves of the brain in carrying out tasks requiring analysis and production of spatial cues. The second experiment's results (also given to 30 controls) showed that Ss with retro-Rolandic lesions performed less well than other Ss. The performance on this kind of task may be affected by damage to the posterior areas of the brain only. The view is supported that posterior corticosubcortical regions are crucial in perception and reproduction of spatial cues and that the contribution of the hemispheres to these functions is not balanced. (21 refs.) - *T. C. Turner.*

University of Milan
Milan, Italy

3046 **BABENKOVA, R. D.; & IUROVSKII, S. IU.** Spatial organization of movements by auxiliary school pupils. *Soviet Education*, 14(1,2,3,): 73-81, 1971-1972.

In a study of 100 MR pupils in grades 1 through 8 of a special school, an attempt was made to determine the capability of each pupil to organize his movements according to given parameters. Most elementary movement tasks were performed on the basis of oral instruction. A series of experiments was set up subsequently to establish the ability of the children to reproduce a given volume of movements precisely. The findings demonstrated that most of the pupils had difficulty in fixing their gaze, in tracking moving objects, and in searching visually for a given object. They had particular difficulty in fixing their gaze on moving fingers. Further analysis of the data showed that in the process of special education, there is a qualitative change in all of the elements that predetermine the possible formation of spatial orientation in MR children, and pupils perform their movements with increasing confidence. During their entire stay in school, MR children continuously improve the precision of their spatial assessment of movements to be performed, and the increase in precision in the performance of movements in 1 spatial direction exerts a positive effect on precision in the performance of movements in another direction. (No refs.) - *B. J. Grylack.*

U.S.S.R. Academy of Pedagogical Sciences
Moscow, U.S.S.R.

3047 **COLLINS, DEAN T.** Head-banging: Its meaning and management in the severely retarded adult. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 220-227.

Head-banging is an exaggeration of the more benign rhythmic activity of body-rocking. A longitudinal study of a 26-year-old male SMR, in whom head-banging persisted into adulthood, confirms head-banging as an extreme manifestation of body-rocking. A treatment program has developed which produced positive results within weeks. Different types of rocking serve distinct purposes in the development of the child. Normative or transient rocking provides reassurance in the face of frustration in mastering a new stage of development. Agitated rocking offers autoerotic gratification in place of maternal deprivation or infant-mother disturbance, and repetitious rocking is an effort to maintain an awareness of the body. This last form of rocking can lead to head-banging, an attempt to differentiate the self from the environment and establish body image. (11 refs.) - *S. Glinsky.*

The Menninger Clinic
Topeka, Kansas

DEVELOPMENTAL ASPECTS – Mental

3048 **GOLDMAN, HERBERT I.; LIEBMAN, O. BERNARD; FREUDENTHAL, ROSLYN; & REUBEN, RICHARD.** Effects of early dietary protein intake on low-birth-weight infants: Evaluation at 3 years of age. *Journal of Pediatrics*, 78(1): 126-129, 1971.

A follow-up study of a group of infants with birth weights below 2,000 g. who were randomly assigned to either a 2 or 4% protein diet has indicated the need for long-term clinical trials to assess the late effects of variation in early nutritional intake. Of the original group of 304 infants, 234 were available for evaluation at approximately 3 yr. The only significant difference in physical abnormalities observed for the 2 groups was excess strabismus in the 4% diet group. Assessment of IQ, utilizing the Stanford-Binet test, form LM, disclosed that scores below 70 occurred with equal frequency in both groups; however, more IQ and CIQ (IQ corrected for weeks of prematurity) scores between 70 and 89 occurred in the 4% diet group, and more CIQ scores above 110 occurred in the 2% diet group. The differences, however, were not statistically significant ($p>0.05$). Two other differences between the 2 diet groups were observed: among those with birthweights of less than 1,300 g. 6 of 26 and 16 of 21 Ss of the 2% and the 4% diet groups, respectively, had IQ's of less than 90 ($p = 0.01$); among undergrown Ss, 9 of 39 and 21 of 45 of the 2% and the 4% diet groups, respectively, had IQ's of less than 90 ($p = 0.05$). (13 refs.) - *M. S. Fish.*

Long Island Jewish Medical Center
New Hyde Park, New York 11040

3049 **BROCKMAN, LOIS M.; & RICCUITI, HENRY N.** Severe protein-calorie malnutrition and cognitive development in infancy and early childhood. *Developmental Psychology*, 4(3): 312-319, 1971.

Twenty severely malnourished Ss (CA11-43 months) and 19 adequately nourished Ss were tested with 10 different sorting tasks to determine the effects of protein-calorie malnutrition during infancy. The experimental Ss showed a lower level of cognitive development on the sorting tasks than did the control Ss. These findings were in accord with previous research showing retardation in social-emotional, social-language and sensory integration areas of development among malnourished children. Relatively permanent cognitive retardation was indicated by the fact that the experimental Ss showed almost no change on their sorting task scores after 12 weeks of nutritional treatment. Experimental Ss also showed smaller head circumference than was normal for age, indicating reduced brain weight and protein content; it is an indication of the severity of nutritional deprivation on actual growth of brain cells. (15 refs.) - *T. C. Turner.*

University of North Dakota
Grand Forks, North Dakota 58201

3050 **BERRY, FRANKLIN M.; JOUBERT, CHARLES E.; & BAUMEISTER, ALFRED A.** Single-letter cue selection and degree of paired-associate learning in retardates. *Journal of Experimental Psychology*, 88(2): 196-204, 1971.

An experiment was conducted to provide information concerning: (a) the point during the course of paired-associate (PA) learning at which retardates begin using single letters as their stimuli; and (b) the effects of overtraining on an already established strategy of selecting single letters. Retarded Ss learned PAs; in this phase each S was assigned to one of 7 groups of PA learning (1,2,3,5,7,2 perfect or 8 overlearning trials), and transfer tests were given following the PA learning stage. It was found that first letters were best recall clues. The notion is therefore supported that retarded Ss select single letters from trigrams as their stimuli for response recall. With increasing degrees of PA learning, two-letter usages are frequently employed. (17 refs.) - *T. C. Turner.*

Columbus College
Columbus, Georgia 31907

3051 **STEPHENS, BETH; & McLAUGHLIN, JOHN A.** Analysis of performances by normals and retardates on Piagetian reasoning assessments as a function of verbal ability. *Perceptual and Motor Skills*, 32(3): 868-870, 1971.

Language was not a significant modifier of performance on 29 Piagetian reasoning assessments. Ss were 75 normals (CA range 6 to 18 yrs; IQ range 90-110; MA range 6 to 19 yrs) and 75 MRs (CA range 6 to 18 yrs; IQ range 50-75; MA range 3 to 13 yrs). When the Wechsler Vocabulary scaled score was used as the covariate to determine the relationship between verbal ability and performance on reasoning tasks, significant differences between normal and MR Ss were found on 29 variables. When Verbal IQ was used as the covariate, 13 of these variables were significant, indicating that verbal ability, as measured by the Wechsler IQ, accounted for some of the variance between normals and MRs. (2 refs.) - *J. K. Wyatt.*

Temple University
Philadelphia, Pennsylvania

3052 **THOMAS, ELIZABETH C.** Conceptual development in advantaged and disadvantaged kindergarten children. *Perceptual and Motor Skills*, 32(3): 711-717, 1971.

White advantaged kindergarten children performed at a higher level than white disadvantaged kindergarten children on an object-sorting test which included spontaneous and structured sorting tasks. The 30 white advantaged Ss (mean IQ 114) were significantly different ($p<.01$) from the 30 white disadvantaged kindergarten Ss (mean IQ 89) in regard to mental age, parents' educational level, occupation of father, restriction of home environment, stimulation in the home, availability of record player, and attendance at movies. Categories of responses by advantaged Ss were 77% classification, 20% early classification, and 3% nonclassification. Disadvantaged Ss' responses were 27% classification, 30% early classification, and 39% nonclassification. Distribution of responses by mental age supported a 5-level hierarchy of conceptual development. Neither sex nor task differences were significant. (11 refs.) - *J. K. Wyatt.*

Children's Medical Center
Tulsa, Oklahoma

3053 **DEICH, RUTH F.** Reading time and error rates for normal and retarded readers. *Perceptual and Motor Skills*, 32(3): 689-690, 1971.

Retarded readers made significantly ($p<.05$) more errors than normal readers but read at the same speed as their reading-grade-level peers on a task which required them to read aloud a series of words which varied in length and familiarity. The performance of 4 groups of Ss was compared: 15 second grade normal readers of average intelligence (mean IQ 107.40, mean WRAT score 3.49); 15 sixth grade retarded readers of average intelligence (mean IQ 100; mean WRAT socre 3.78): 15 bright second graders (mean IQ 131, mean WRAT score 5.76); and 10 sixth grade retarded readers (mean IQ 95.20, mean WRAT score 6.35). Within-group comparisons indicated that sixth grade normal readers with higher scores on the Wide Range Achievement Test (WRAT) read significantly faster ($p<.01$) and made significantly fewer errors ($p<.01$) on 3- and 6-letter familiar words than second grade normal readers with lower WRAT scores. (4 refs.) - *J. K. Wyatt.*

Pacific State Hospital
Pomona, California

3054 **MONTROY, PHERYL; *McMANIS, DONALD; & BELL, DONALD.** Development of time concepts in normal and retarded children. *Psychological Reports*, 28(3): 895-902, 1971.

Tests measuring concepts of time—simultaneity, equality of synchronous intervals, and order of events—administered to 36 normal and retarded children matched for MA demonstrated that for all Ss the active ordering of events was the most difficult. A sequential order of time-concept development was suggested by the greater time—concept understanding associated with MA increases. Essential similarity of understanding simultaneity, equality of synchronous intervals, and active ordering of events was noted at higher MAs; at lower MAs, children showed greater understanding of the first 2 items. Although normals seemed to display a faster developmental rate than did retardates, the confusion of IQ with MA level in the retardates suggested that final judgment be withheld. (4 refs.) - *B. Berman.*

*Eastern Washington State College
Cheney, Washington 99004

3055 **JOESTING, JOAN; & *JOESTING, ROBERT.** Correlation of scores on the Picture Interpretation Test and Stanford-Binet Form L-M IQs. *Psychological Reports*, 28(3): 906, 1971.

Correlation of scores on the Picture Interpretation Test (PIT) and the Stanford-Binet Form L-M Test, administered to 27 Ss (CA 4.9 to 13.8 years) referred for behavior problems, demonstrated that (1) skills of learning to read a picture are apparently learned through looking at pictures in books, asking questions, and receiving encouragement all of which are, in turn, related to the child's intelligence; and (2) children's IQs may be increased through exposure to suitable educational stimulation. Such programs for disadvantaged children particularly could be implemented in day-care centers. (4 refs.) - *B. Berman.*

*Challenge Foundation
Chapel Hill, North Carolina 27514

3056 **KAUFFMAN, JAMES M.; PAYNE, JAMES S.; & ENSMINGER, E. EUGENE.** Quantitative judgments of culturally advantaged and disadvantaged preschool children. *Psychological Reports*, 28(3): 939-944, 1971.

A comparison of cognitive functioning in 35 culturally advantaged and 35 disadvantaged children (CA 2 yrs, 4 mos to 4 yrs, 7 mos) showed that conservation-like cognitive functions (concrete intellectual operations) are influenced by discrimination-task difficulty, cultural disadvantages, and age of advantaged Ss. Psycholinguistic and motivational variables, which are known to contribute significantly to the apparent cognitive deficits of disadvantaged children, were minimized. Most of the differential responses to spatial orientation occurred among the 3 youngest groups. Contrary to reports by Mehler and Bever (1967), children as young as 2 yrs, 4 mos did not exhibit any form of "conservation" behavior, and "non-conservation" does not seem to be a temporary cognitive characteristic of children attributable to misleading perceptual strategies. (14 refs.) - *B. Berman.*

University of Virginia
Charlottesville, Virginia

3057 **GOLDEN, MARK; BIRNS, BEVERLY; BRIDGER, WAGNER; & MOSS, ABIGAIL.** Social-class differentiation in cognitive development among Black preschool children. *Child Development*, 42(1): 37-45, 1971.

Eighty-nine Black children from different social classes were tested on the Stanford-Binet at 3 years of age as part of a longitudinal follow-up study of children who had been tested on the Cattell Infant Intelligence Scale at 18 and 24 months of age. The study was made to determine whether the same pattern of social-class differentiation in cognitive development that was reported to emerge during the third year of life in White children was also present in Black children. Although there were no significant social-class differences on the Cattell at 18 and 24 months, highly significant socioeconomic status (SES) differences in intellectual performance were revealed when the same children were tested on the Stanford-Binet at age 3. The range in mean IQs of the Black children in the extreme SES groups was 93 to 116. When such factors as birth complications, poor health, and nutrition are excluded, it appears that social class differences in intellectual performance have not been demonstrated until the third year of life. Infant tests such as the Cattell, which measure primarily perceptual-motor skills, may not be sufficiently sensitive to detect the differences during the first 2 years of life. (12 refs.) - *B. J. Grylack.*

Albert Einstein College of Medicine
of the Yeshiva University
Bronx, New York 10461

3058 **GUINAGH, BARRY J.** An experimental study of basic learning ability and intelligence in low-socioeconomic-status children. *Child Development*, 42(1): 27-36, 1971.

Forty low socioeconomic status (SES) Black children and 40 low-SES White children were used in a study conducted to determine the effect of a training program on scores of Raven's Progressive Matrices (RPM), a test of intelligence. It was predicted that children with high basic learning ability, as measured by a test of digit span (DS), would improve more than children with low DS scores. An analysis of variance using

an RPM pretest, posttest, and retention test as repeated measures showed the main effect due to treatment to be statistically reliable ($F=36.9$, $p<.001$). In the low-SES white group, both the high and low DS Ss scored significantly better on the RPM posttest than the respective control groups. In the low-SES Black group, only the high DS experimental group scored better on the RPM posttest than its control group. While the results of the study brought into serious question assertions made about the genetic immutability of abstract abilities, the functional dependence of the abstract abilities tested by the RPM on basic learning ability was equivocal. (7 refs.) - *B. J. Grylack.*

University of Florida
Institute for Development of Human Resources
Gainesville, Florida 32601

3059 CAFFREY, BERNARD; JONES, JUDITH D.; & HINKLE, BARBARA R. Variability in reaction times of normal and educable mentally retarded children. *Perceptual and Motor Skills*, 32(1): 255-258, 1971.

A comparison of 10 normal and 10 EMR children on differences and variabilities in reaction time showed the EMR group to be slower in both simple and choice tasks. They were also more variable than normals on the Lafayette visual choice reaction-time test. It is suggested that a simple task for the MR was equivalent to a choice task for normals and that factors in addition to cognition may be part of the MR response. (6 refs.) - *E. L. Rowan.*

Clemson University
Clemson, South Carolina 29631

3060 STRICKLAND, STEPHEN P. Can slum children learn? *American Education*, 7(6): 3-7, 1971.

MR is not necessarily perpetuated in families. The interim results of the Milwaukee Project, currently in its fifth year, seem to refute family perpetuation of MR. Forty mothers of newborn infants are participating, all with an IQ of less than 70. Two-thirds of the infants were enrolled in an experimental program, while the remaining third functioned as a control group. A carefully structured educational and activities program was made available to the experimental group, the oldest children of which are now approaching their fifth birthdays. At 42 months of age, the children in this group were rated at an average of 33 intelligence quotient points higher than the control group, with some registering as high as 135. (No refs.) - *S. Glinsky.*

U.S. Office of Education
Washington, D.C. 20202

3061 HAGEN, JOHN W.; & HUNTSMAN, NANCY J. Selective attention in mental retardates. *Developmental Psychology*, 5(1): 151-160, 1971.

The performance of normal and MR children was compared in an investigation of the significance of selective attention in discrimination learning tasks. In the first of 2 studies, 47 pupils in educable and trainable special classes for MRs, 14 children in a private nursery school, and 47 pupils attending normal public school classes were compared on performance in the central versus incidental task recall paradigm. Four MA levels were included. A single significant interaction effect, MA X Central-Incidental, was observed. There was no distinction between normal and MR Ss in their ability to attend selectively when matched for MA. The results confirmed that central versus incidental task performance showed the same improvement with increasing MA as has been found with CA. The second study duplicated the first but used 21 institutionalized MR Ss at 2 MA levels. There was no significant difference among the 2 institutionalized groups as compared with the data of previously tested normal and MR Ss ($F<1.00$). However, a significant MA effect ($F=7.44$, $df=1/75$, $p<.01$) was found, and interaction between groups and central-incidental task performance approached significance. The data indicate that the institutionalized Ss differed from the other groups in the ability to attend selectively. It is questionable whether institutionalization in itself is the only cause of the deficit. (12 refs.) - *B. J. Grylack.*

University of Michigan
Ann Arbor, Michigan 48104

3062 BROWN, ANN L. Response latencies for intradimensional and extradimensional

transfer of the oddity solution. *Psychonomic Science*, 23(6): 389-391, 1971.

Extradimensional transfers (ED) of the oddity solution gave rise to longer response latencies than did intradimensional transfers (ID). Twenty institutionalized MRs were assigned randomly to 4 conditions comparing ID and ED oddity shifts in either a within or between Ss embedded problems design. Following extended training the MRs demonstrated high accuracy of responding for all shift conditions. ED shifts of the vehicle dimension for oddity resulted in significantly longer latencies than either ID or control shifts. The disruptive effects of ED shifts were found only when the transfer problems contained variable irrelevant dimensions and when comparison was made between groups which received either ID or ED shifts but not both conditions. (9 refs.) - *S. Glinsky.*

University of Connecticut
Storrs, Connecticut 06268

3063 **HARTER, SUSAN; BROWN, LYNN; & ZIGLER, EDWARD.** The discrimination learning of normal and retarded children as a function of penalty conditions and etiology of the retarded. *Child Development*, 42(2): 517-536, 1971.

Ss were MA matched normal, familial retarded, and organically retarded children who were given a size-discrimination task under Reward Only condition (correct choice reinforced) and Reward + Penalty condition (correct choice reinforced, incorrect choice penalized). Normal and familial MR Ss showed better learning in Reward + Penalty condition, while organics did not. The former groups used strategy behavior, while the latter displayed position preference behavior. It is suggested that etiology be used as a dimension in the design of studies examining normal-retarded differences in behavior. (32 refs.) - *T. C. Turner.*

Yale University
New Haven, Connecticut

3064 **BERG, NORMAN L.; & BERG, SANDRA D.** Comparison of verbal intelligence of young children from low and middle socioeconomic status. *Psychological Reports*, 28(2): 559-562, 1971.

Twenty Ss from a low socioeconomic status (SES) school district and 20 Ss from a middle SES school district were tested by the same examiner during a 2-week period to determine the relationship between language and socioeconomic level. Ss were tested for verbal intelligence by measurement of their hearing vocabulary as an aspect of language. The results do not support the hypothesis that young children of middle SES score higher in verbal intelligence on the Peabody Picture Vocabulary Test (Peabody) than young children of low SES. The finding of a significant main effect ($F=4.91$, $p<.05$) for race indicated that Blacks scored significantly lower on Peabody IQ than did Whites in both school districts. Although the hypothesis was not confirmed, analysis of data demonstrating that overall racial differences do exist is considered to be an important finding. The fact that white children scored higher on Peabody IQ indicates that differential programs and remedial help within schools undergoing integration might be advisable. (8 refs.) - *B. J. Grylack.*

Xavier University
Cincinnati, Ohio

3065 **DAS, J. P.; & BOWER, A. C.** Orientating responses of mentally retarded and normal subjects to word-signals. *British Journal of Psychology*, 62(1): 89-96, 1971.

MRs exhibited a higher level of galvanic skin responses (GSRS) to nonsignal words during a vigilance task than did normals. The GSRS of normal and MR children of comparable CA were recorded during a 30 minute vigilance task consisting of 2 signal and 4 nonsignal words. One of the signal words served as a warning signal, and the second served as an imperative signal for button pressing. The warning signal evoked a greater number of GSRS in the normals than the imperative one, and this sequence was reversed in the MRs. GSR adaptation to signal words occurred at the same rate and level for both groups. In adaptation to the nonsignal words the MRs maintained a higher level of GSR frequencies than the normals. These results were not congruent with an attentional deficit model of retardation. (15 refs.) - *S. Glinsky.*

University of Alberta
Edmonton, Canada

3066 **PISHKIN, VLADIMIR; & BURN, JULIAN.** Concept identification in the brain-damaged: Intertrial interval and information complexity. *Journal of Abnormal Psychology*, 77(2): 205-210, 1971.

Brain-damaged individuals were variably affected by information complexity and intertrial interval in a concept identification (CI) problem. The 90 Ss were divided into a brain-damaged and a control group. The CI performance of the control Ss was optimally facilitated at the 15 second interval, but was not affected in the brain-damaged Ss. There was a linear increase in CI errors as a function of complexity in both groups, although brain-damaged Ss demonstrated a deficit on CI in terms of decision time and processing of available information. (19 refs.) - *S. Glinsky.*

Veterans Administration Hospital
Oklahoma City, Oklahoma 73104

3067 **GOLDSTEIN, HARRIS S.; & PECK, ROSALIND.** Cognitive functions in Negro and White children in a child guidance clinic. *Psychological Reports*, 28(2): 379-384, 1971.

In order to determine whether racial differences exist in cognitive functioning of children seeking help from a mental health clinic, the cognitive abilities of Negro and White children attending an inner-city child guidance clinic were examined with the Wechsler Intelligence Scale for Children (WISC) and the Harris modification of the Goodenough Draw-A-Man test. Ss were 22 White males, 34 Negro males, 18 White females, and 17 Negro females. White males scored significantly higher ($p<.001$) than Negro males only on the verbal part of the WISC. White females achieved significantly higher scores ($p<.001$) than Negro females on verbal, performance, and full scale IQs as well as on the Harris scores. There was no significant difference between White and Negro males on the Harris scores. From the standpoint of intellectual functioning, Negro and White boys are equally good candidates for psychotherapy. The differences between the 2 groups of girls were complicated by marked differences in the personality problems exhibited. (6 refs.) - *B. J. Grylack.*

Downstate Medical Center
Brooklyn, New York 11203

3068 **GREER, BOBBY G.; & WHITLEY, CAROLYN.** The relationship between birth order and learning disabilities. *Exceptional Children*, 37(8): 608, 1971.

This exploratory study's hypothesis was that the larger the family and the later the child's birth order, the greater was the chance for a child to have a learning disability. A previous pilot study (Psychological Services Division of the Memphis City Schools) was the derivation for the hypothesis. Records of 200 learning disabled Ss were chosen randomly from the Special Education Division of the Memphis City Schools. Age, sex, birth order, and number of children in the family were recorded for each S. Ss came from families ranging in size from 1-9 children, with 3.21 being the average number of children per family. A significant proportion of Ss came from large families and, although little can be concluded from the study, it opens up a new area for investigation: why children with learning disabilities do tend to come from larger families. (No refs.) - *T. C. Turner.*

Memphis State University
Dept. of Special Education and Rehabilitation
Memphis, Tennessee

3069 **BAUMEISTER, ALFRED A.; & CAMPBELL, CECIL.** Formation of backward associations in paired-associates learning by normal children and retardates. *Journal of Experimental Psychology*, 89(2): 298-305, 1971.

Two experiments on bidirectional learning, utilizing free stimulus and response learning with normal and retarded children, demonstrated that forward learning is only modestly superior to backward learning. In the first experiment (64 normals and 64 retardates: mean IQ 69) groups of Ss were stopped at different points in forward paired-associates (PA) learning and then tested for backward associations; in the second, Ss were compared on both unidirectional and bidirectional items. During PA learning, backward-association strength was constant, but, as shown by conditional probability analysis, PA learning in normals and retardates did not show complete associative symmetry. Both groups manifested effective spontaneous learning in both the background and the bidirectional associations. (12 refs.) - *B. Berman.*

University of Alabama
University, Alabama

3070 **CLARKE, A.D.B.** Stretching their learning skills. *Journal of Rehabilitation in Asia*, 12(3): 5-11, 1971.

Learning abilities of the mentally handicapped have been grossly underestimated—MRs need more complex learning experiences, for they have capabilities never before suspected. Research has shown retardate deficits can be modified and ameliorated. It is not in their best interests to reduce demands on them and simplify their lives. As the retarded child becomes more independent physically and meets new problems he must be shown how to generalize from previous experiences and to transfer to new problems. Experiments with "transfer tasks" have shown this generalization process does occur. Practice in one form of categorization improves ability to undertake other forms. Studies using Thorndike-Lorge word lists have demonstrated unsuspected memory capabilities in MRs. Appropriate training will help the mentally handicapped cope with difficult new situations requiring intelligent organization. (12 refs.) - *B. Berman.*

No address

3071 **SCHWARZ, ROBERT H.; & COOK, JOHN J.** Mental age as a predictor of academic achievement. *Education and Training of the Mentally Retarded*, 6(1):12-15, 1971.

The relationship of mental age (MA) to academic achievement among culturally disadvantaged MR children was investigated. Level of academic achievement for 499 MR students (CA 74-184 months, IQ 51-82) in a disadvantaged rural population was determined by the Wide Range Achievement Test (an average of the reading and arithmetic subtest scores). In each case this level was compared to the expected achievement in terms of MA as reflected by the test manual. Ss were also divided in terms of CA and grouped in 10-point increments of IQ, and IQ groups were compared in terms of relationship of MA to achievement level. Differences between expected and actual academic achievement increased as a function of age at all the IQ levels (51-60, 61-70, 71-80). It was concluded that factors other than MA alone must influence achievement and that, while MA may be a reasonably valid measure of developmental level, its use as a standard for educational placement is questionable. (16 refs.) - *K. L. Woodworth.*

University of Wisconsin
Madison, Wisconsin

3072 **GORDON, DONALD A.; & BAUMEISTER, ALFRED A.** The use of verbal mediation in the retarded as a function of developmental level and response availability. *Journal of Experimental Child Psychology*, 12(1): 95-105, 1971.

One hundred forty-four male and female Ss were selected from a state residential institution for MRs for participation in a study of the use of verbal mediation in the retarded as a function of developmental level and response availability. Three levels of MA and 3 instructional conditions were 2 between-subjects variables. Recognition versus recall task for paired-associate learning was a within-subjects variable. Instructions, MA level, and response availability all showed significant effects as variables. The results supported a developmental interpretation of mediational learning processes in MRs. In general, the results did not support claims that retardates cannot generate or employ their own mediations. Compared to standard paired-associate instructions, instructions either to generate their own or to use examiner-supplied mediators produced superior performances for all retardates, regardless of IQ or MA. (12 refs.) - *B. J. Grylack.*

University of Oregon Medical School
Portland, Oregon

3073 **BUFFORD, RODGER K.** Discrimination and instructions as factors in the control of nonreinforced imitation. *Journal of Experimental Child Psychology*, 12(1): 35-50, 1971.

Three experiments were conducted to evaluate the variables which determine performance of verbal nonreinforced imitation. In the first experiment, discriminability was increased through systematic reduction of the number of responses in the reinforced class. Based upon the activity of 6 children aged 4.9 to 7.2 years, of whom 4 were MR, the most striking finding was the

consistently high level of nonreinforced response. However, the function of discrimination in these responses was equivocal. The second and third experiments were designed to evaluate the influence of verbal instructions on nonreinforced responses. When not instructed specifically to do otherwise, Ss consistently followed the SAY instructions which had been given only during the first session of the experiments. However, despite a much longer history of responding in the absence of SAY instructions, the effect of the DON'T SAY instruction was to reduce, temporarily, nonreinforced imitation. It appears that instructions function as setting events whose effects perish over extended periods even when not repeated. (13 refs.) - *B. J. Grylack.*

University of Illinois
Champaign-Urbana, Illinois

3074 **RASKIN, LARRY M.; FONG, LOUELLA J.; & BLACK, KATHRYN N.** Perceptual recognition and guessing behavior by normal and educable retarded children. *Psychonomic Science,* 22(6): 371-372, 1971.

EMRs showed lower levels of perceptual recognition and greater levels of guessing behavior than normals. Normal and EMR children at 2 MA levels were tested on recognition of fragmented or incomplete stimulus forms. A series of 12 slides was presented, and the number of brief exposure periods required to make a correct identification was recorded. The results indicated that the younger MA Ss needed more exposures to identify the forms. Although there were no IQ effects, the EMRs guessed incorrectly more often than the normals, and the younger EMRs guessed more than all other children. (7 refs.) - *S. Glinsky.*

Purdue University
Lafayette, Indiana 47907

3075 **JENSEN, ARTHUR R.** The role of verbal mediation in mental development. *Journal of Genetic Psychology,* 118 (First half): 39-70, 1971.

Much of what comprises learning and intelligence involves various verbal and symbolic mediational processes, and theory and research pertaining to verbal mediation focuses on (1) the nature of mental development, and (2) social concern with the educational problems of the "culturally disadvantaged." Both "growth readiness" and "cumulative learning" models reflect recent research, but cumulative learning is most in tune with potential for educating the culturally disadvantaged. Mediation theory seeks to comprehend the forms of behavior classed as "thinking": types of mediation processes include labeling, mediated and semantic generalization, far transposition, reversal-nonreversal shift, experimentally acquired mediation, extraexperimentally acquired mediators, associative clustering, verbal self-reinforcement, syntactical mediation and mnemonic elaboration, learning set formation, and cross-modal transfer. Recent research has emphasized stimulation and development of cognitive processes in deficient children. Research problems include arrangement of learning processes, training cognitive skills, associative clustering in free recall, Piaget conservation tests, figure coyping, mediation arousal, and relationship between learning abilities and intelligence. (49 refs.) - *B. Berman.*

University of California
Berkeley, California

3076 **CLARKE, BRYAN R.; & LESLIE, PERRY T.** Visual-motor skills and reading ability of deaf children. *Perceptual and Motor Skills,* 33(1): 263-268, 1971.

Tests of visual-motor skills and reading ability were administered to 59 deaf children aged 8 to 12 years in order to improve methods of detecting very retarded readers among the deaf at an early age and to recommend areas of research in remediation. On the basis of performance IQ and results of the Gates-MacGinitie Reading Tests, 3 comparable groups of 9 children were selected. In Group I, reading was retarded by more than 2 years, and performance IQs were less than 90. Group II reading was retarded by more than 2 years, and performance IQs were greater than 90. Reading was retarded by less than 2 years, and performance IQs were greater than 90 in Group III. Results showed a significant difference between Groups I and II, but no significant difference was found between the average profile of Groups I and II combined and the Group III profile. Retarded and nonretarded deaf readers

were not differentiated by the tests used. Ongoing research should be directed towards acquisition of language and the relation of language to the reading skills of deaf children. (22 refs.) - *B. J. Grylack.*

University of British Columbia
Vancouver, Canada

3077 **SPITZ, HERMAN H.; & WEBRECK, CINDY A.** Effects of age and mental retardation on temporal integration of visual stimuli. *Perceptual and Motor Skills,* 33(1): 3-10, 1971.

Thirty-two normal eighth-graders were compared with 32 normal third-graders and 32 retardates with a mean IQ of 62 in an investigation of the effects of CA and MR on the temporal integration of visual stimuli. Ten pairs of visual stimuli were used in a way that each half of a pair would appear as meaningless segmented lines until the 2 halves were fused and revealed a drawing of an animal. Analysis of variance indicated no significant groups effect, a significant interstimulus interval (ISI) effect ($p<.001$), and no significant interaction. However, the groups were fairly disparate at 10- and 40-millisecond ISIs. No other ISI differences at any points were significant. The prediction that lower MA groups would perform better than higher MA groups was not substantiated. Sensory traces, assumed to exist for lower MA Ss on the basis of threshold for dark interval of longer duration and experiments on backward masking, did not produce the effect expected in a temporal integration experiment. (18 refs.) - *B. J. Grylack.*

E. R. Johnstone Training and Research Center
Bordentown, New Jersey

3078 **SHOTICK, ANDREW; & BLUE, MILTON.** Influence of CA and IQ levels on structure and amount of spontaneous verbalization. *Psychological Reports,* 29(1): 275-281, 1971.

Ninety-six children ranging in CA from 4 years 0 months to 11 years 11 months were Ss in an investigation of the use of spoken syntactical structures and amount of spontaneous verbalization of children of varying levels of intelligence (retarded, average, and superior) at 4 CA levels. Intelligence levels were established with the Stanford-Binet Test of Intelligence, Form LM, and spontaneous verbalization was elicited with a series of 10 individually mounted photographs of everyday situations. Four classes of expression units were designated as Complete Communication Unit (CUc), Incomplete Communication Unit (CUi), Partial, and Maze, depending upon the bases of semantic and grammatical correctness. An analysis of variance of total word output scores showed no significant differences among intellectual levels or CA groups and no interaction. CA 8 to 9 accounted for the greatest increase in the use of CUc for both average and retarded Ss, whereas superior children showed their greatest forward movement at CA 6 to 7. It appears that the use of semantically complete language forms has been established by age 4 or 5. The major change occurring after that time lies in the refinement of grammatical usage in oral language. (13 refs.) - *B. J. Grylack.*

University of Georgia
Athens, Georgia

3079 **BROOKS, CLARENCE R.; & CLAIR, THEODORE NAT.** Relationships among visual figure-ground perception, word recognition, IQ, and chronological age. *Perceptual and Motor Skills,* 33(1): 59-62, 1971.

A study of visual figure-ground perception and its relationship to word recognition was made with 118 EMR children aged 7 through 12 years in an attempt to isolate a skill specifically related to the reading process. Analysis of variance indicated a significant difference between readers and nonreaders ($F=9.96$, $df=1/19$, $p<.05$) on visual figure-ground perception. The higher IQ groups appeared to possess better visual figure-ground ability. Age differences showed that visual figure-ground perception is related to reading at the readiness and first reader levels. As EMR children mature, visual figure-ground perception ability is not so important to the reading process and will not differentiate reading-ability groups. The relationship found between visual figure-ground perception and reading indicates a need for research on the use of visual figure-ground training procedures with children who cannot read. Since visual figure-ground perception will distinguish MR readers from nonreaders, it may

be possible to train a child in this skill, after which he would be able to begin reading. (12 refs.) - *B. J. Grylack.*

University of Iowa
Iowa City, Iowa

3080 ROY, R. K. Reminiscence in retardates on a paired-associate task. *Journal of Mental Deficiency Research*, 15(1): 43-50, 1971.

Ss in a 24-hour delayed recall group produced significantly ($p<.01$) more new responses on recall trials than Ss in a 2-minute immediate recall group. Ss were 40 institutionalized MRs (mean IQ 65.3; mean CA 25.5 yrs) who were selected on the basis of a pre-training performance criterion. The task involved recall of a list of paired-associate meaningful materials after 1 reinforced anticipatory trial and 2 nonreinforced trials where the stimuli were presented alone without the paired responses. The relationships between intelligence and short term recall ($p<.05$) and recognition ($p<.05$), block design and short term recall ($p<.01$) and recognition ($p<.05$), and digit span and 24-hour recall and short term recognition ($p<.05$) were inverse. There were significant and positive relationships between intelligence and 24-hour recall ($p<.01$) and recognition ($p<.01$), block design and 24-hour recall ($p<.05$), and digit span and short-term recall ($p<.01$) and 24-hour recognition ($p<.05$). (17 refs.) - *J. K. Wyatt.*

St. Lawrence's Hospital
Caterham, England

3081 HILL, A. LEWIS; & BURKE, DEBORAH. Apparent visual size as a function of age, intelligence, and a surrounding frame of reference for normal and mentally retarded subjects. *Developmental Psychology*, 5(2): 349-356, 1971.

Size constancy and the effect of a surrounding frame of reference on the perception of size were investigated as a function of CA and MA in 68 normal Ss aged 4 to 14 years and 45 MR Ss aged 9 to 20 years (mean IQ=55.21). Size matches increased with age ($r=.57$, $p<.01$) for normal Ss between the ages of 4 and 10. Size matches for normal Ss over 11 years were close to constancy (M=3.83 inches), with little, if any, further development. Size matches were close to constancy in MR Ss at all ages, suggesting that size constancy development was not a function of MA. The effect of the frame of reference decreased slightly for normal Ss when other objects in the room could be seen. The age at which the distance control condition size matches were equal to size matches in constancy control condition was 14 years for MR Ss as compared with 10 years for normal Ss. It appeared that either the mechanisms involved in size matches were influenced both by CA and IQ, or that different mechanisms were involved in those 2 conditions and that those mechanisms developed at different rates for normal and retarded Ss. (16 refs.) - *B. J. Grylack.*

New York State Institute for
Basic Research in Mental Retardation
Staten Island, New York 10314

3082 REIMANIS, GUNARS; & GREEN, RUSSEL F. Imminence of death and intellectual decrement in the aging. *Developmental Psychology*, 5(2): 270-272, 1971.

One hundred and eighty-seven male residents of a Veterans Administration domiciliary participated in a study of differences in the rate of intellectual decline between individuals who died within a year after the last testing and those who died later or were still living. Depending upon which measure the Ss had received previously, either the Wechsler-Bellevue Intelligence Scale or the Wechsler Adult Intelligence Scale was administered. Significant ($p<.05$) differences were found between the group which died within a year and the survivor group on all 3 intelligence measures that were tabulated (Verbal, Performance, and Full Scale). The group which died showed the larger intellectual decrement. The data supported the hypothesis that a significantly greater intellectual decline takes place for a person who is likely to die within a year. However, it could not be shown that the entire decline occurs during the last year. The decline in cognitive functioning with age appears to be due to factors other than time passage, such as physical accidents, disuse of cortical functions, and changes in motivation. (2 refs.) - *B. J. Grylack.*

Corning Community College
Corning, New York 14830

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3083 **CROXEN, MARY E.; & *LYTTON, HUGH.** Reading disability and difficulties in finger localization and right-left discrimination. *Developmental Psychology*, 5(2): 256-262, 1971.

The relationships between finger localization, right-left discrimination, and reading ability were studied in a normal population of 59 boys and 23 girls, 9 to 10 years of age, who had reading quotients of 80 or less and in control Ss of the same age with reading quotients over 80. For boys and girls separately, as well as for the sexes combined, the "average" readers were found to have significantly higher total finger localization scores than the poorer readers. The poorer readers had significantly lower scores on right-left discrimination. Correlations between finger localization scores and right-left discrimination scores showed a small but significant association, even when nonverbal IQ was partialled out, and this correlation was reinforced by the connection between low scores on the finger localization test and total reversal in the right-left discrimination tests. The results were consistent with some previous research which suggested that reading disability is associated with a general perceptual deficit, certainly for some children and more markedly for the poorest readers. (10 refs.) - *B. J. Grylack.*

*University of Calgary
Calgary, Alberta, Canada

3084 **WEI, TAM T. D.; LAVATELLI, CELIA B.; & JONES, R. STEWART.** Piaget's concept of classification: A comparative study of socially disadvantaged and middle-class young children. *Child Development*, 42(3): 919-927, 1971.

Four of Piaget's classification tasks were used in testing 80 Ss from kindergarten and second grade in order to determine any differences in classificatory performances among children of middle-class (MC) and culturally deprived (CD) environments. The tasks were the shifting of criteria, the use of real objects in classification, the inclusion problem, and multiplicative classification. Results supported the hypothesis that both kindergarten and second-grade children of the CD groups would score lower on tasks of classification than children of the MC groups. An analysis of variance showed a significant difference between the 2 social classes ($F=49.62$, $df=1, 76$, $p<.001$). There was no increase in differences between social classes as CA increased. Differences between sexes were not found. Differences in the reasoning process of the 2 social groups were revealed for both age levels. The study supports Piaget's theory of a sequence in logical development and his theory of equilibration, which stresses the continual interaction between the individual and the environment. (8 refs.) - *B. J. Grylack.*

2007 Rainbow View Drive
Urbana, Illinois 61801

3085 **MINUCHIN, PATRICIA.** Correlates of curiosity and exploratory behavior in preschool disadvantaged children. *Child Development*, 42(3): 939-950, 1971.

Observations were made of 18 Negro children in a Head Start preschool setting who ranged in age from 3 years 9 months to 4 years 8 months in order to develop consistent measures of curiosity and exploration which would be applicable to the young in this setting and to investigate the relationship between variations in exploratory behavior and other aspects of emotional and cognitive growth. Sources of data included observations of new experiences in the preschool setting, teacher and observer rankings, and 4 individual sessions with each child. The data suggested that exploratory behavior could be assessed reliably in young children and that ongoing preschool programs could provide reliable data on spontaneous responses to naturalistic situations. Evidence also indicated reasonable consistency in a child's stance towards environmental opportunities, seen along an approach-avoidance continuum. The data could not establish connections between the children's responses and variations in family background, but research along these lines was indicated. (21 refs.) - *B. J. Grylack.*

Temple University
Philadelphia, Pennsylvania 19122

3086 **CHISSOM, BRAD S.** A factor-analytic study of the relationship of motor factors

to academic criteria for first- and third-grade boys. *Child Development*, 42(4): 1133-1143, 1971.

An examination of the motor characteristics of development as related to achievement in school or any change in the structure of motor abilities in the developmental process was made in order to define more thoroughly the motor relationship to academic achievement and aptitude. Ss were 169 boys who were given tests of balance, dynamic strength, and gross motor co-ordination. The Otis-Lennon Measure of Academic Aptitude was also given. A teacher rating measure of academic achievement was obtained for each child. Results indicated that motor factor structure for boys in both grades was the same. Data support the relationship between motor factors of balance and motor coordination and academic achievement and aptitude. Perceptual-motor theories are more applicable for boys at earlier ages. The relationship between certain motor abilities and intellectual abilities is not supported for older boys. (28 refs.) - *T. C. Turner.*

Georgia Southern College
Statesboro, Georgia 30458

3087 BECKWITH, LEILA. Relationships between attributes of mothers and their infants' IQ scores. *Child Development*, 42(4): 1083-1097, 1971.

Relationships among maternal and infant behaviors and individual differences in IQ scores of 24 adoptive, family reared white middle class infants are investigated. Two visits were made to the 24 infants' homes by 2 observers; at each visit the Cattell Infant Intelligence Scale and motor items from the Gesell Developmental Schedules were given. Mother and baby were observed for 1 hour in spontaneous interaction and activities. Observational categories were: verbalizations to observers, verbal and physical contacts with infant, ignoring infant's cry and/or social approaches, permissiveness in allowing infant to explore objects, places, own capabilities and body. Preceding the second session each mother answered the Schaefer-Bell Parental Attitude Response Inventory. No relationship was found between postnatal socioeconomic class membership and infants' intellectual development. Specific maternal behaviors affected infants' gross motor and intellectual performances. Babies' scores on the Cattell test seemed to be enhanced by increased social experience outside home and parents. Both the Cattell and Gesell scores were affected by either prenatal nutrition or genetic factors indexed by natural mother socioeconomic class. (27 refs.) - *T. C. Turner.*

760 Westwood Plaza
Los Angeles, California 90024

3088 TSVETKOVA, L. S. Concerning methods for examining children with speech impairments. *Soviet Education*, 14(1,2,3,): 157-165, 1971-1972.

Fifty normally developed children of different preschool ages, 50 children with MR and various forms of alalia and dyslalia, and 10 children with MR were studied to determine the differences among them with regard to the visual perception of objects and the stability of visual conceptions. Among normal Ss, there was an increase in the volume of memorization as a function of age. Characteristics of the Ss with retarded speech development consisted of a considerable decline in the general volume and precision of memorization as compared with normal Ss as well as a very pronounced inertia of conceptions. Verbalization of the subject world can occur only after the development of a perceptual basis for speech. Analysis of the genesis of children's speech shows that it develops in close interaction with processes of acoustical, visual, visuospatial, and somatospatial perception and with motor responses. The examination of the child should culminate in a general assessment of the psychopathological picture of the impairment and should indicate the nature and possible mechanisms of speech impairment. (No refs.) - *B. J. Grylack.*

Moscow M. V. Lomonosov State University
Moscow, U.S.S.R.

DEVELOPMENTAL ASPECTS – Social and Emotional

3089 **FAY, WARREN H.** On normal and autistic pronouns. *Journal of Speech and Hearing Disorders*, 36(2): 242-249, 1971.

Pronominal reversals in the autistic child are analyzed both in terms of ego theory and a contradictory grammatical approach emanating from studies of childhood echolalia. Ego-based approaches have failed to recognize the problems all children face in meeting personal pronouns and have rested on inferences concerning presumed failure in self-resolution, while ignoring linguistic problems presented by shifting personal pronouns. The grammatical approach regards psycholinguistic confusion to be a primary problem in the acquisition process. The focus of treatment is shifted from the "I" (with which the child has little acquaintance) to developing the you/me dichotomy, and from expression to comprehension. (31 refs.) - *T. C. Turner.*

University of Oregon Medical School
Portland, Oregon

3090 **MacMILLAN, DONALD L.; & KEOGH, BARBARA K.** Normal and retarded children's expectancy for failure. *Developmental Psychology*, 4(3): 343-348, 1971.

Sixty Ss in special education classes for the EMR and 60 Ss in regular third grade classes were interviewed individually and asked to manipulate 9 red and white Kohs blocks to duplicate designs on 5x8 inch cards. Three designs were completed and 3 interrupted before completion. A repetition choice was allowed and the choice noted; Ss were then asked why some of the tasks had not been completed. EMR and normal Ss perceived interruption differently, the former seeing it as a personal failure, while the latter did not. It is concluded that the expectancy for failure is characteristic of the problem-solving approach of the retarded child. (12 refs.) - *T. C. Turner.*

University of California
Riverside, California 92502

3091 **OPPEL, WALLACE C.; & ROYSTON, ANITA B.** Teen-age births: Some social, psychological, and physical sequelae. *American Journal of Public Health*, 61(4): 751-756, 1971.

A study of teen-age mothers suggests that deficits in the child's physical, social, and/or psychological development seemed related to the youth of the mother. In a 12-year longitudinal study, 86 mothers under 18 years of age were compared with 86 mothers 18 years and older. Mothers under age 18 were less likely to remain with their children or to rear them in a healthy family atmosphere. They were more apt to increase their family during the 6 to 8 years after the birth of the child. They displayed less anxiety about their children and were more apt to believe the children should be given the freedom to behave independently. Incidence of infantile behavior problems or acting out behavior during examination by a psychologist was higher for children of younger mothers. These children were more likely to be deficient in reading-grade level, lower in IQ, underweight, and shorter. They were more likely to be described as outgoing, dependent, and distractible. (9 refs.) - *J. K. Wyatt.*

Johns Hopkins University
School of Hygiene and Public Health
Baltimore, Maryland 21205

3092 **EGG, MARIA.** Les enfants deficients mentaux grandissent aussi! (Mentally deficient children grow also!) *Nos Enfants Inadaptes*, 37 (first trimester): 12-15, 1971.

In the majority of mentally deficient children, physical as well as mental development is affected. Contrary to popular conceptions, however, the sexual desire of mental deficients is no stronger than that of normal youth. In fact, sex plays a less important role in the lives of MR children, among whom autostimulation is prevalent. It is during the period of mental maturity

that the future course of life of MRs is decided. Acknowledgement of these individuals is insufficient to integrate them into society. They deserve to work and to receive a salary, and their confidence in themselves is greatly dependent upon their self-achievement. In addition, entrance into the salaried society symbolizes their admission into the social structure of society and their acceptance as human beings. (No refs.) - *B. J. Grylack.*

No address

3093 **YANDO, REGINA; & *ZIGLER, EDWARD.** Outerdirectedness in the problem-solving of institutionalized and noninstitutionalized normal and retarded children. *Developmental Psychology*, 4(2): 277-288, 1971.

Outerdirectedness, or the reliance on external cues rather than on one's own cognitive resources, is characteristic of problem solving in MR children. Eight equal groups (N=24) of institutionalized and noninstitutionalized children with organic MR, familial MR, normal intelligence matched by CA, and normal intelligence matched by MA were given a task of 3 choice discriminations in which a light went on above a wrong choice and a task entailing imitation of designs made by an experimenter or a machine. The MR children made significantly more cued than noncued errors and were more imitative than normals. Younger children imitated more than older children, but institutionalized normal children tended to reject cues from adults. The institutionalized organics were more outer-directed than institutionalized familials. Outer-directedness apparently reflects early experience in trying to please adults as well as the child's intellectual level. (9 refs.) - *E. L. Rowan.*

*Yale University
New Haven, Connecticut 06510

3094 **FIEDLER, MIRIAM F.; LENNEBERG, ERIC H.; ROLFE, URSULA T.; & DRORBAUGH, JAMES E.** A speech screening procedure with three-year-old children. *Pediatrics*, 48(2): 268-276, 1971.

Forty-six children who failed a speech screening examination for 3-year-olds were compared to 92 control Ss matched for age, sex, and time of examination, who had passed the screening test, in an attempt to determine causes of speech delay. Significant differences were found between the 2 groups of children in complications of pregnancy and labor (.05 level), prematurity (.01 level), abnormal cry at birth (.01 level), delayed first year development (.001 level), atypical first year behavior (.05 level), unfavorable environment (.01 level), and occurrence of hospital procedures in the first year of life (.01 level). Psychological testing of the same children at 4 years of age and psychological and neurological examinations of these Ss at 7 years of age revealed the persistence of significant differences between the 2 groups in the occurrence of various psychological and neurological deviations from the normal. (13 refs.) - *B. J. Grylack.*

Maternal and Infant Health Study
Boston, Massachusetts 02115

3095 **FOREHAND, REX; & BAUMEISTER, ALFRED A.** Stereotyped body rocking as a function of situation, IQ, and time. *Journal of Clinical Psychology*, 27(3): 324-326, 1971.

Recordings of stereotyped body-rocking behavior in 12 male institutionalized MR (6 had untestable IQs; 6 had IQs of 21 to 31, with varying verbal, self-help, and socialization skills) showed a higher stereotypy rate for high IQ MRs in a restricting environment; location did not affect the rate for lower IQs. Discrepancies between the two groups resulted probably from variations in responsiveness to environmental stimuli. Body rockers apparently are not a homogeneous group with respect to situational changes. (9 refs.) - *B. Berman.*

University of Alabama
University, Alabama

3096 **FOREHAND, REX; & BAUMEISTER, ALFRED A.** Rate of stereotyped body rocking of severe retardates as a function of frustration of goal-directed behavior. *Journal of Abnormal Psychology*, 78(1): 35-42, 1971.

SMRs showed an increased rate of stereotyped body rocking in response to frustration of goal-directed behavior. The Ss were SMRs divided

into 3 groups of body rockers. Two groups differed on the basis of the number of reinforcements received for lever pulling prior to frustration and the third served as a control for changes in rate of stereotypy over time. Frustration was defined as interference with goal-directed activity. After a prefrustration measure of stereotypy was obtained, the 2 experimental groups were exposed to a frustration situation on each of 4 days in which goal-directed lever pulling was blocked. Stereotypy rate and activity level were recorded during prefrustration and frustration periods. The results indicated that frustration was associated with increased rate of stereotypy, which was directly related to the number of reinforcements prior to the blocking of goal-directed behavior. (12 refs.) - *S. Glinsky.*

University of Alabama
Box 6302, University, Alabama 35486

3097 **BALLINGER, BRIAN R.** Minor self-injury. *British Journal of Psychiatry*, 118(546): 535-538, 1971.

A survey of minor self-injury in patients in an MR hospital and psychiatric hospital revealed a considerable incidence of self-injury. In 1 month 93 out of 626 MRs and 20 out of 584 psychiatric patients had injured themselves. Self-injury bears a certain relationship to other characteristics of the patients. Individual attention, recreation, and constructive occupations may be more important than drug treatment in the alleviation of these symptoms. (6 refs.) - *S. Glinsky.*

Royal Dundee Liff Hospital
Liff, Dundee, Scotland

3098 **SILVERMAN, MITCHELL.** Developmental trends in the vocational interests of special education and normal students. *Journal of Genetic Psychology*, 118 (Second half): 157-172, 1971.

A comparison of developing vocational interests in 45 normals and 45 retardates showed, despite their varying educational backgrounds, similar trends in development of vocational interests and an absence of significant group-by-age interactions. The groups did, however, show significant differences in preference for adventuresome occupations (retardates made consistently fewer responses) and in jobs representing low-level mechanical skill (retardates made significantly more responses). These differences, it was assumed, reflected differential early treatment, the effects of being tagged as retarded. In factors showing age effects, both groups revealed general to selective response trends. Retardates showed backwardness in developing vocational interests in jobs reflecting low-level mechanical skill, mechanical and quantitative skills, and production in terms of changing, molding, and manipulating the environment. A pictorial measure of occupations listed in Holland's models was used to assess vocational interests. (7 refs.) - *B. Berman.*

University of South Florida
Tampa, Florida

3099 **FLOOR, LUCRETIA; *ROSEN, MARVIN; BAXTER, DONALD; HOROWITZ, JUDITH; & WEBER, CINDA.** Socio-sexual problems in mentally handicapped females. *American Institute for Mental Studies Training School Bulletin*, 68(2): 106-112, 1971.

The possible relations between sex-related infractions occurring within the institution and socio-sexual problems occurring in the community is investigated. Ss were 49 females who had been discharged from Elwyn Institute between 1963 and 1970 and returned to independent community life after completing rehabilitation programs. Data were obtained from continuing community evaluations of Elwyn "graduates" as part of a longitudinal follow-up study and from medical and psychiatric reports, houseparent notations, and educational information. Ss were divided into "High Problem and Low Problem" groups. The "High Problem" defining criterion was that the individual's sexual activity in the community had resulted in severe economic, employment, or personal difficulties to the point of dysfunction and that the individual must have sought help from some outside source. "High Problem" women tended to be younger than "Low Problem" women. A search of the records was performed to examine the relationship between sex-related infractions at Elwyn and later community problems ($X^2=7.6$; $p<.01$). Ss with severe community problems were also likely to have had histories of sex-related infractions within the institution. Ss demonstrating such behaviors may

be showing their need for appropriate counseling, structured learning experiences, and support. Appropriate co-education and socialization experiences should therefore be an important part of community preparation for institution residents. (2 refs.) - *T. C. Turner.*

Elwyn Institute
Elwyn, Pennsylvania

3100 **EDMONSON, BARBARA; & CEGELKA, WALTER J.** Parent-school collaboration via educational merit badges? *American Institute for Mental Studies Training School Bulletin*, 68(2): 97-105, 1971.

Three stresses of adolescence--discontinuity in training, separation of youth from adult world during years of formal education, and membership in a culture with goals and values unlike the adult culture--are explored and related to the MR. Parents of EMR, although abandoning hope of a high-level vocation, should not give up expectations of responsible independence. To foster a sense of competence in children, parents should encourage them to undertake tasks that seem to contribute to family life. Successful continuity of training is a matter of deciding on small steps of responsible behavior and keeping them clearly in mind so that progress is acknowledged. Parents can do much to equip the MR child with essential social skills. Windows and bridges to the adult world can be provided when parents and teachers collaborate. In considering the social problems of lack of clarity with respect to goals and to what is accomplished and failure to receive social recognition for achievement, the merit badge plan has been offered as a possible answer. Clarity is provided by the clearly defined, measurable tasks in a merit badge manual, and recognition is provided by the badge awarded for each area of competence. To transfer this device to education would require that educational goals of personal, social, economic, and civic adequacy be translated into sets of clearly defined tasks, which would be described in a manual available to pupils. The pupils would then select their competency targets and be coached by family and friends during practice sessions. (25 refs.) - *T. C. Turner.*

University of Oregon
Eugene, Oregon

3101 **SMEETS, PAUL M.** Some characteristics of mental defectives displaying self-mutilative behaviors. *American Institute for Mental Studies Training School Bulletin*, 68(2): 131-135, 1971.

Four hundred MR and emotionally disturbed Ss were surveyed in a study to determine: (a) the number of Ss with self-mutilative behavior (SMB); (b) whether there were differences relative to age; (c) the age at which SMB was first noticed; (d) the relationship between IQ and SMB; (e) the relationship between SMB and hyperactive, stereotyped, aggressive, destructive, and unharmful tantrum behaviors. Of the 400 Ss, 35 showed 1 or more forms of SMB. Of the 12 types of SMB found, 6 were variations of injury to the head. The group with severe SMB comprised almost 3 times as many males as females, but no overall relationship between sex difference and intensity of mutilation could be established (Phi. 05), nor was age related to the severity of SMB. Case histories which included reference to the onset of SMB were available in 14 cases and provided the information that SMB appeared before the age of 4 in 50 percent of those cases and after the age of 4 in 50 percent. These figures disagree with most case studies on SMB, which suggest that the period between 15 months and 4 years is most critical for its onset. There appears to be positive relationship between a decrease in IQ and the severity of SMB, but cases were found to occur at various levels of intellectual functioning. The results also suggest that individuals who severely mutilate themselves are, as a rule, more maladjusted than those who engage in mild SMB. (15 refs.) - *T. C. Turner.*

The Woods Schools
Langhorne, Pennsylvania

3102 **PAYNE, JAMES E.** The effect of institutionalization on educable mentally retardates' expectancy of failure. *American Institute for Mental Studies Training School Bulletin*, 68(2): 77-81, 1971.

A study was undertaken to investigate the hypothesis that EMR would show a decrease in expectancy of failure following placement in an institution oriented to a more SMR norm. Ss were 60 EMR (CA10-20 years; IQ 50 or above). They were chosen on the basis of residence in the institution as experimental variable: 30 short-

term (ST) Ss (6 months or less residency) and 30 long-term (LT) Ss (12-24 months residency). Reactions to failure of ST and LT group Ss were compared. Ss were first asked to perform a sorting task, failure was then induced on another task at which Ss were working for a prize, and finally Ss repeated the initial task. Data were analyzed primarily by comparing differences in change scores. Change in performance was determined by net raw score change and magnitude and directionality of change. The differences were in the predicted direction, but none were significant at the .05 level of confidence. A possible explanation for this was in the definition of 6 months or less ST placement, which had been done from research necessity. But with an ST residence strictly defined, the data would support the conclusion that MR children experience a reduction in failure expectancy. (14 refs.) - *T. C. Turner.*

University of Texas at Austin
Austin, Texas

3103 **NORMAN, ARTHUR; & MELVILLE, CHARLES H.** Potential applications of telemetered heart rate data to developmental psychology. *Developmental Psychology*, 5(2): 190-194, 1971.

Two children participated in a demonstration of the instrumentation, procedures, and some potential research applications involved in the recording of heart rate by telemetry. The electrodes which recorded the beats were connected to a direct modulated FM transmitter, attached to a polygraph displaying each heart beat. The highest heart rate in the 4-year-old normal girl was recorded when she was called by name by an examiner and smiled. The highest heart rate in the 9-year-old retarded boy was obtained while he leaned back in his chair after touching his pants zipper. Evidence has shown that heart rate deceleration is related to "attention," "acceptance of external information," or efforts to increase environmental intake, while heart rate acceleration is found during types of problem solving and during "rejection of sensory stimulation." Biotelemetry may be useful in the investigation of cognitive functioning in psychotic as well as normal and retarded Ss and in the employment of physiological changes to infer psychological processes that might never have been considered if overt behavior alone were observed. (19 refs.) - *B. J. Grylack.*

Georgia Mental Health Institute
Atlanta, Georgia 30306

3104 **SEVERY, LAWRENCE J.; & DAVIS, KEITH E.** Helping behavior among normal and retarded children. *Child Development*, 42(4): 1017-1031, 1971.

The use of distinctions in helping behavior (conceptualization, development issues, mental status and social skills, research settings and design limitations) is shown to be facilitative in dealing with important issues in the development of social skills and prosocial behavior. Ss were retarded and normal (CA 3 to 5 and 8 to 10 years) children who were observed and coded regarding trait ratings, milieu descriptions, psychological versus task helping, attempted versus achieved help, and retarded sample helping. Total helping behavior data showed younger normal and older MR Ss to be more helpful than older normal and younger MR Ss. There is a need to distinguish between showing concern and actual helping behavior and between attempting to help and succeeding in helping, since the conceptual importance of the distinction lies in its relevance to the difference between motivation and competence. The tentative explanation for the failure of older normals to observe greater helpfulness is that, while their capacities to help and recognize the need for help have grown, they have also been socialized into other norms serving to inhibit helping among same-age peers, primary among these norms being achievement and independence norms feeding competitiveness. MR children have not developed such stress on these norms and therefore acquired inhibitions do not hide the greater competence of older retardates. (16 refs.) - *T. C. Turner.*

University of Florida Department of Psychology
Gainesville, Florida

3105 **SHALIMOV, V. F.** Composite studies of oligophrenics. *Soviet Education*, 14(1,2,3): 49-61, 1971-1972.

The degree of social, occupational, and family adaptation of 78 persons afflicted with oligophrenia who graduated from a special school for

the handicapped between 1935 and 1956 was established in a study of their composite profiles. Ss were divided into 4 groups. Thirty-one were diagnosed to have the basic variant of the disease, in which the underdevelopment of the most complex forms of cognitive activity was not accompanied by severe damage of 1 or another sense organ or by impairment of the emotional or volitional sphere. The underdevelopment of cognitive activity was accompanied by neurodynamic impairment in 32 Ss and by behavior resembling that of psychopaths in 9, and it was combined with gross focal disturbances in 6. The level of social, occupational, and family adaptation was found to be dependent upon the structure of the defect of the oligophrenic, with Ss suffering from the basic variant demonstrating the most favorable adapation and Ss with oligophrenia complicated by psychopathological behavior manifesting the least successful adaptation. (No refs.) - *B. J. Grylack.*

U.S.S.R. Academy of Pedagogical Sciences
Moscow, U.S.S.R.

3106 **MENOLASCINO, FRANK J.** Psychiatric aspects of mongoloidism. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 241-249.

A high percentage of emotional disturbance in mongoloids casts doubt on the image of the mongoloid as immune to psychiatric disturbance. The Ss were 86 mongoloids studied by a multidisciplinary team between 1958-1963. Eleven of the Ss presented with 3 general types of emotional disturbances and family configurations. Fifty per cent of these displayed abnormal electroencephalograms, while only 13.4% of the remaining 75 mongoloids exhibited such abnormalities. This finding suggests that cerebral dysrhythmia may be etiologically significant in producing emotional disturbances in these children. Much more attention must be given to the psychiatric aspects of mongoloidism. (32 refs.) - *S. Glinsky.*

University of Nebraska College of Medicine
Omaha, Nebraska

3107 **WEBSTER, THOMAS G.** Problems in emotional development in young retarded children. In: Menolascino, Frank J. ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 28-37.

MR is a clinical developmental syndrome which regularly includes impairment in emotional as well as in intellectual development. The Ss were 159 preschool MRs with every type of MR and ranged in IQ from 20 to 85. Diagnostic studies were made with a focus upon disturbances in emotional development. None of the MRs exhibited an emotional development comparable to a nonretarded child of the same MA, with 35% showing mild, 48% moderate, and 17% severe disturbance. The primary psychopathology of the MR syndrome was described particularly in terms of the development disturbance and special descriptive traits, such as nonpsychotic autism, repetitiousness, inflexibility, passivity and flexibility in emotional life. Varying importance attaches to the factors of organic brain damage, validity of the sample findings for the overall MR population, and the effect of deprivation on emotional disturbance. (17 refs.) - *S. Glinsky.*

National Institute of Mental Health
Division of Manpower and Training Programs
Chevy Chase, Maryland

DEVELOPMENTAL ASPECTS – Psychodiagnostics

3108 **WEINER, PAUL S.** Stability and validity of two measures of intelligence used with children whose language development is delayed. *Journal of Speech and Hearing Research,* 14(2): 254-261, 1971.

Data are presented on the validity and reliability of the Arthur Adaptation of the Leiter International Performance Scale (AALIPS) and the Peabody Picture Vocabulary Test (PPVT) in assessing the intelligence of preschool children

with delayed language capacities. Tests were given 3 times at several-month intervals, the Weschler Intelligence Scale for Children (WISC) also being given at the last testing session. The mean AALIPS 92 did not change significantly on retesting, remaining at a low average level. Neither did the PPVT 92, which stayed within the dull normal range. Changes in individual scores were considerable on both tests. By comparing the last testing results of the AALIPS and PPVT with the last results on the WISC, concurrent validity was explored. It was concluded that AALIPS is a highly useful, reliable and valid testing measure, while there is some doubt about the PPVT. In both the AALIPS and PPVT, results are stable if precise 92's are used for studies of groups, but are not so useful for studies of individuals. The AALIPS results show stability over time and can predict later functioning on an intelligence test. The PPVT has definite limitations as a measure of adequacy of verbal functioning. It best measures the comprehension of single words, but has less applicability as a measure of intelligence than AALIPS. (11 refs.) - *T. C. Turner.*

University of Chicago
Chicago, Illinois

3109 **ERTL, JOHN P.** Fourier analysis of evoked potentials and human intelligence. *Nature*, 230(5295): 525-526, 1971.

Intelligence as measured by IQ tests shows no correlation with characteristics of average evoked potential. From an FM library of a total of more than 1,000 tapes was selected 2 groups of Ss: 93 high IQ Ss with scores of over 120 on 2 of 3 tests, and 71 low IQ Ss having scores less than 85 on 2 out of 3 tests (Otis, PMA, and WISC). Mean ages were 11 and 11.6 years for the high and low IQ groups, respectively; of the total, 92 were male and 72 were female. Fourier analysis of EEG data derived from bipolar scalp electrodes showed no significant differences between the average Fourier transforms of the 2 groups at average evoked potential of either 4.5 s or 4.25 s after stimulation. (9 refs.) - *M. S. Fish.*

University of Ottawa
Ottawa, Canada

3110 **GUNDERSON, BERNICE V.** Diagnosis of learning disabilities—the team approach. *Journal of Learning Disabilities*, 4(2): 107-113, 1971.

The contribution of each professional to the complex field of learning disabilities is outlined and an inventory of areas to assess and instruments or techniques to provide an overall profile of a child's strengths or weaknesses is presented. Communication and cooperation among disciplines is seen as the key to success in treating a child's learning problems. (11 refs.) - *T. C. Turner.*

SW Minnesota State College
Marshall, Minnesota

3111 **JOHNSON, DALE L.; & JOHNSON, CARMEN A.** Comparison of four intelligence tests used with culturally disadvantaged children. *Psychological Reports*, 28(1): 209-210, 1971.

Twenty-nine 5-year-old Head Start children were given the Slosson Intelligence Test (SIT), the Peabody Picture Vocabulary Test (PPVT), and the Draw-A-Man Test (DAMT) and these scores were compared with those from the Revised Stanford-Binet (S-B). The most striking feature of the results was that while the SIT, PPVT, and DAMT were all fairly well correlated with the much longer S-B, the obtained mean IQs varied greatly. The SIT is a brief, useful, valid measure of young children's intelligence, since it correlated .79 with the S-B, and its mean IQ was almost the same as the S-B mean. (8 refs.) - *T. C. Turner.*

University of Houston
Houston, Texas

3112 **COSTELLO, JOAN; & ALI, FAIZUNISA.** Reliability and validity of Peabody Picture Vocabulary Test scores of disadvantaged preschool children. *Psychological Reports*, 28(3): 755-760, 1971.

The standard Peabody Picture Vocabulary Test (PPVT) administered to 36 Black preschoolers yielded test-retest correlations of 0.77; a modified version of the PPVT produced values of 0.87 and 0.80. Correlation of the standard version with 2 other measures of intellectual traits and with teacher ratings of various verbal characteristics yielded a fair validity, with the notable

exception of grammatical accuracy. PPVT scores would appear to have adequate stability over a brief time period with groups of preschool disadvantaged children. PPVT, while it does not measure the same characteristics as other individual tests, does share some variance in common. However, PPVT scores should not be considered alone as adequate for either intellectual or language evaluation. (10 refs.) - *B. Berman.*

Yale Child Study Center
New Haven, Connecticut

3112 **TIGAY, BARRY; & KEMPLER, HYMAN L.** Stability of WISC scores of children hospitalized for emotional disturbance. *Perceptual and Motor Skills*, 32(2): 487-490, 1971.

Records of 25 hospitalized emotionally disturbed children (CA 7 yrs, 5 mos to 12 yrs, 9 mos at first testing) tested on the WISC at least twice during hospitalization and once after hospitalization showed the test to be a reliable instrument for measuring intelligence in emotionally disturbed youngsters. Except for the Comprehension subtest, reliability coefficients and mean differences demonstrated as high or higher stability on verbal subtests than those reported for non-disturbed Ss. Test-retest studies showed that, except for Picture Arrangement, which is more reliable than test-retest data for normals, performance-subtest stability is significantly lower for disturbed Ss. On retesting, both performance and full-scale IQs showed a marked increase. (8 refs.) - *B.Berman.*

Wayne State University
Detroit, Michigan

3114 **WOO-SAM, JAMES; ZIMMERMAN, IRLA L.; & ROGAL, RICHARD.** Location of injury and Wechsler indices of mental deterioration. *Perceptual and Motor Skills*, 32(2): 407-411, 1971.

Administration of 4 indices of mental deterioration from the Wechsler-Bellevue Test to 95 head–injured veterans, classified on the basis of head injury and homogeneous in age and IQ, revealed significant differences among the indices for occipital, frontal, and parietal groups, but not for the mixed frontal and parietal groups. Except for the Wechsler Revised index, efficiency of the indices was not affected by location of injury. Generalized concentration impairment is a characteristic of head injury, and it is measured by poor performance on the Digit Symbol subtest. Diagnostic impairments show certain impairments that are syndrome-specific and play a key part in differential diagnosis. In brain injury, it appears that the locus of injury is associated with differential impairments readily measured by the Wechsler test. (15 refs.) - *B. Berman.*

Rancho Los Amigos Hospital
Downey, California

3115 **JOHNSON, JAMES E.; HELLKAMP, DAVID T.; & LOTTMAN, THOMAS J.** The relationship between intelligence, brain damage, and Hutt-Briskin errors on the Bender-Gestalt. *Journal of Clinical Psychology*, 27(1): 84-85, 1971.

The Hutt and Briskin scoring system for the Bender-Gestalt reflects both organicity and intelligence. Matched groups of 20 organic and 20 nonorganic state hospital patients were divided into 6 different intelligence levels as measured by WAIS IQ scores. The Hutt-Briskin scoring system used on their Bender drawings was able to diagnose the majority of organic subjects in the IQ range of 60 to 89. The presence of organicity (i.e. more than 5 errors), then, can be determined from the Bender-Gestalt only when the subject is of borderline or dull-normal intelligence. Nonorganic subjects or organic subjects of normal intelligence do not make many errors on the test. (6 refs.) - *E. L. Rowan.*

Loyola University
Chicago, Illinois

3116 **ALI, FAIZUNISA; & COSTELLO, JOAN.** Modification of the Peabody Picture Vocabulary Test. *Developmental Psychology*, 5(1): 86-91, 1971.

Improvement in the administration of the Peabody Picture Vocabulary Test(PPVT) was sought by modifying those aspects of the test which appear to affect adversely the performance of disadvantaged preschool children. The major areas of modification were expectancy (test

length and ceiling), reinforcement (style and schedule), and specification of instructions. Ss were 2 groups of disadvantaged Black children attending 2 nursery schools. In order to determine test-retest stability of each form, the PPVT was administered twice with a 2-week interval, and the modified form (MPPVT) was administered twice 6 months later, with the same test-retest interval. Test-retest reliability (Pearson r) for the MPPVT was .86 as compared to .77 for the PPVT. Mean PPVT score was 29.0, and mean MPPVT score, 36.0 ($t=3.65$, $p<.002$). Median scores for the PPVT and MPPVT were 29.5 and 37.0, respectively. No significant sex difference in performance was noted for either test administration, although in both instances male mean scores were slightly higher than female mean scores. Both males and females performed better on the MPPVT. The results indicate that the MPPVT positively influences test scores and is well accepted by examiners. (23 refs.) - *B. J. Grylack.*

Institute for Juvenile Research
Chicago, Illinois 60611

3117 **FRANKENBURG, WILLIAM K.; CAMP, BONNIE W.; & VAN NATTA, P. A.** Validity of the Denver Developmental Screening Test. *Child Development,* 42(2): 475-485, 1971.

Validity of the Denver Developmental Screening Test (DDST) was tested by collecting systematic data concerning its relationship with other tests. Ss were 236 children who were administered the DDST, Stanford-Binet, Revised Yale Developmental Schedule (RYDS), Cattell Infant Intelligence Scale, and Bayley Infant Scale Tests. The analysis of MA received from scoring the DDST is in general agreement with results from the criterion tests. Strong support is lent to the use of DDST as a screening device for identifying developmental deviations. (10 refs.) - *T. C. Turner.*

University of Colorado Medical Center
Denver, Colorado

3118 **GOFFENEY, BARBARA; HENDERSON, NORMAN B.; & BUTLER, BRUCE V.** Negro-White, male-female eight-month developmental scores compared with seven-year WISC and Bender test scores. *Child Development,* 42(2): 595-604, 1971.

The effectiveness of the Bayley Infant Scales in predicting 7-year Wisc and Bender test scores for Negro and White, male and female Ss is reported. Ss were 626 children whose scores were correlated. Findings were: (1) all 8-month measures for the total population correlated significantly with all 7-year measures; (2) no 8-month score for the Negro male group correlated significantly with any later IQ or Bender score; (3) the 8-month mental and fine motor scores for each female group correlated significantly with full and verbal scale IQ; mental scores also correlated significantly with performance IQ and Bender. The study points out the usefulness of a single test to predict the effectiveness of later test scores, but the authors are cautiously optimistic about the predictive value of infant scales. (13 refs.) - *T. C. Turner.*

No address

3119 **PASEWARK, RICHARD A.; FITZGERALD, BERNARD J.; & GLOECKLER, TED.** Relationship of Peabody Picture Vocabulary Test and Wechsler Intelligence Scale for Children in an educable retarded group: A cautionary note. *Psychological Reports,* 28(2): 405-406, 1971.

Forty-nine EMR students ranging in age from 7 to 14 years were tested to determine the relationship between the Peabody Picture Vocabulary Test (Peabody) and the Wechsler Intelligence Scale for Children (WISC) as used in evaluating MR. It was found that the Peabody IQ consistently overestimates WISC IQs. Intercorrelations between IQs from these 2 tests were poor. Although reduced correlations might be anticipated because of the restricted intellectual range of the Ss, their low magnitudes warn against use of the Peabody to designate the level of retardation of a particular individual. (3 refs.) - *B. J. Grylack.*

University of Wyoming
Laramie, Wyoming

3120 **ABLES, BILLIE S.** The use of the Draw-A-Man Test with borderline retarded children without pronounced pathology. *Journal of Clinical Psychology*, 27(2):262-263, 1971.

The relationship between the Draw-a-Man (DAM), Binet, and WISC measures of intelligence on 3 varying populations is explored. Records were those of 74 children 14 or under who had full scores on the WISC or Binet and had done a full drawing of a man. It is concluded that the DAM can be used with reasonable confidence as an intelligence measurement of children who are being tested primarily for intellectual assessment and who do not have organic and/or emotional problems. (2 refs.) - *T. C. Turner.*

University of Kentucky Medical School
Lexington, Kentucky

3121 **DANIELS, LLOYD K.; & STEWART, JAMES A.** Development of a vocational adjustment rating scale for the mentally retarded. *American Institute for Mental Studies Training School Bulletin*, 68(1): 10-14, 1971.

The standardization of a vocational ajustment rating scale for MRs and a description of the methods used to establish reliability and validity are discussed. The scale uses the checklist rating method and has 4 response categories; its final form consists of 2 separate measures of work adjustment: 23 items considered as indirectly and 39 items considered as directly related to work success. The true factor composition of the scales of direct/indirect traits of vocational adjustment and the predictive validity of the scale still require empirical justification. (5 refs.) - *T. C. Turner.*

Central Connecticut College
New Britain, Connecticut

3122 **SIMPSON, C. DENE; & VEGA, ARTHUR.** Unilateral brain damage and patterns of age-corrected WAIS subtest scores. *Journal of Clinical Psychology*, 27(2):204-206, 1971.

Left vs. right discriminations of cerebral damage were made possible by comparing WAIS and subtest scores of unilateral damage groups with those of a control group. Age-corrected scaled scores were used to avoid confusion between performance deficit associated with increasing age and that resulting from cerebral disorder. Ss were 88 patients with brain damage; 50 Ss without brain damage were controls. The WAIS was given to each patient as part of the Halstead-Reitan Neuropsychological Test Battery. Brain-damaged groups all showed impairment and were lower in intellectual functioning than the controls. The possibility of using the results of the test as a screening test for the presence of unilateral brain damage is seen as promising. (17 refs.) - *T. C. Turner.*

Northwest Nazarene College
Nampa, Idaho 83651

3123 **OBRZUT, JOHN E.; & THWEATT, ROGER C.** An investigation of the Sherman Mental Impairment Test: A normative study. *Journal of Clinical Psychology*, 27(2): 224-225, 1971.

A validation study of the Sherman Mental Impairment Test (SMIT) is reported which extends Sherman's findings to another norm group not included in the original study. Ss were 50 brain-injured persons and 50 normals. The SMIT was individually given and scored for Letter Finding Score (LFS) and Reaction Time Score (RTS). Brain-damaged Ss significantly differed from the controls. Several males and females were misclassified when just RTS were used, suggesting that the test does not discriminate sex differences. (4 refs.) - *T. C. Turner.*

No address

3124 **PUSTEL, G.; STERNLICHT, M.; & DEUTSCH, M.** Feminine tendencies in figure drawings by male homosexual retarded dyads. *Journal of Clinical Psychology*, 27(2): 260-261, 1971.

An exploratory effort to test the validity of existing sets of criteria as indices of the presence or absence of femininity in figure drawings by MR male homosexuals in an institution is reported. Ss were 18 pairs (dyads) of male homosexuals, each dyad consisting of a "mas-

culine" (active) and "feminine" (passive) partner, who drew an adult male human figure, a picture of himself, and a picture of an animal he chose. The Ss drew figures of a human male characterized by a strong quality of femininity; the tendency was more pronounced among the passive members of the dyads. The passive Ss also produced a greater number of physically small and nonaggressive animals than did the active members. (3 refs.) - *T. C. Turner.*

Willowbrook State School
Staten Island, New York

3125 **ISRAEL, LAWRENCE J.; &*HEAL, LAIRD W.** A developmental study of the subject matter choices in the free drawing of trainable mentally retarded subjects. *Exceptional Children*, 37(8): 597-600, 1971.

Trainable mentally retarded (TMR) Ss (25 boys and 19 girls, MA2-6 years, CA8-14 years) drew from 20 to over 100 drawings apiece. Two judges tabulated human schema details in 14 categories (hair, ears, hands, for example), and the results are presented in tables. Significant differences were found in subject matter preferences of TMR children and nonretarded children of comparable MA. TMR's drew more humans and fewer weapons, instruments, and ships than nonretarded children; higher MA children drew more plants than humans. The investigators concluded from their study that art samples can be reliably categorized and that their subject matter is definitely related to the intellectual development of the artist. This may be helpful in planning art programs for the TMR. (2 refs.) - *T. C. Turner.*

*George Peabody College
Nashville, Tennessee

3126 **VIRSU, VEIJO; & ELONEN, ANNA S.** Reproduction of superimposed forms from dot patterns in five diagnostic groups of children. *Journal of Clinical Psychology*, 27(3): 345-350, 1971.

Significant differences in drawing geometric patterns were displayed by 151 Finnish children (divided into 5 diagnostic groups: MR, entirely disturbed, convulsive, brain-damaged, normal) tested with the KTK Dot Pattern Test. In 7 variables (Corrections, Rotation, Disproportion, Impotence, Concreteness, Incompletion, Number of Items Drawn) the groups showed noticeable variations when IQ and age were partialled out: emotionally disturbed, brain-damaged, and normals were most successfully classified. None of the 16 drawing variables, however, was dominantly characteristic of any specific diagnostic group, although rotation of figures and concreteness occurred most often with the brain-damaged. Classification of MRs was at chance level. A fairly high percentage of correct classifications showed the advantages of a multidimensional test. (10 refs.) - *B. Berman.*

University of Michigan
Ann Arbor, Michigan

3127 **HASAZI, JOSEPH E.; ALLEN, ROBERT M.; & WOHLFORD, PAUL.** Effects of mode of administration on Bender-Gestalt Test performance of familial retardates. *Journal of Clinical Psychology*, 27(3): 360-362, 1971.

Administration of the Bender-Gestalt Test (under 2 conditions: (a) standard and (b) each design on a separate card identical with the design card) to 40 MR showed fewer rotation, perseveration, and fragmentation errors when designs were reproduced on separate cards. A decrease in perseveration errors may have resulted from a decrease in size of the reproduction field; decline in fragmentation errors may have been caused by the increase in reproduction fields. Performance aspects considered diagnostic are partly a function of test-administration factors. (6 refs.) - *B. Berman.*

University of Vermont
Burlington, Vermont

3128 **MITCHELL, MARLYS M.** Analysis of Welsh Figure Preference Test scores of educable mentally handicapped children. *American Institute for Mental Studies Training School Bulletin*, 67(4): 214-219, 1971.

Administration of the Revised Art Scale (RA) of the Welsh Figure Preference Test (WFPT) to 183

EMR, 48 normal second-grade children, and 47 normal sixth-grade children showed that EMRs scored significantly lower than normal sixth graders but not significantly lower than the second graders. Significant sex differences on the RA scale were found among EMRs (boys scored significantly higher) but not among normals: there were no significant differences on the don't like (DL) scale. Mean DL scores of the sixth-grade group differed significantly from those of the EMR group. Results suggest further research on sex differences of EMR children and on differences between the RA and DL scales. The tests seem useful in studying differences between EMR children and other groups. (6 refs.) - *B. Berman.*

University of North Carolina
Chapel Hill, North Carolina

3129 **DeWOLFE, ALAN S.; BARRELL, ROBERT P.; BECKER, BRUCE C.; & SPANER, FRED E.** Intellectual deficit in chronic schizophrenia and brain damage. *Journal of Consulting and Clinical Psychology*, 36(2): 197-204, 1971.

Patterns of intellectual deficit independent of group mean levels of performance were compared using 50 chronic schizophrenics and 50 Ss with nonlateralized brain damage. In each category, Ss were divided into two groups: those 26-59 years, and those over 60 years of age. The two groups at each age level were matched on age and education. Administered were all 11 subtests of the Wechsler Adult Intelligence Scale (WAIS) and 9 subtests from the Halstead Battery of Neuropsychological Measures. Results showed significant interaction of WAIS subtests with diagnosis and a statistically reliable interaction of Halstead Battery subtests with diagnosis and age. Other pattern analyses showed that the younger schizophrenics and brain damaged Ss differed in WAIS Comprehension and Digit Span and in Halstead Battery Category. The schizophrenic and brain damaged Ss over 60 differed in WAIS Comprehension, Digit Span, Picture Completion, Block Design, and Halstead Battery Speech Discrimination. Results suggest that the pattern of intellectual deficit in chronic schizophrenia differs from the pattern for nonlateralized brain damage. (16 refs.) - *K. L. Woodworth.*

Veterans Administration Hospital
Downey, Illinois 60064

3130 **RESNICK, ROBERT J.; & ENTIN, ALAN D.** Is an abbreviated form of the WISC valid for Afro-American children? *Journal of Consulting and Clinical Psychology*, 36(1): 97-99, 1971.

Although full and abbreviated forms of the Wechsler Intelligence Scale for Children (WISC) show statistically highly significant correlations, comparison of mean differences between the forms (administered to 80 urban Afro-American children) yielded information contradicting this implied validity, and showed changes in diagnostic IQ level with the shortened form. In verbal IQ, 30 Ss showed changes in intelligence category, 23 spuriously showing more ability on the shortened form. Changes of 5 or more points were shown by 35, 41, and 34 Ss on verbal IQ, performance IQ, and full-scale IQ, respectively. These discrepancies in intelligence levels question the validity of the shortened form when used with Black children, and, indeed, with White children and adults as well. (8 refs.) - *B. Berman.*

Medical College of Virginia
Richmond, Virginia

3131 **WATSON, CHARLES G.** An MMPI scale to separate brain-damaged from schizophrenic men. *Journal of Consulting and Clinical Psychology*, 36(1): 121-125, 1971.

Three MMPI (Minnesota Multiphasic Personality Inventory) scales have been developed that successfully differentiate male organics (brain-damaged) from schizophrenics, but fail to differentiate among female populations. The 3 scales (a) unweighted form of 80 items differentially endorsed at a Veterans Administration psychiatric hospital; (b) the same items weighted as a function of their differentiating power; and (c) a 30-item form weighted in accordance with the items' differentiating strength, were cross-validated in 2 hospital populations. Results of the scales when used with females were predictable, since female psychiatric patients show different behavior patterns from males. The scales should not be used as general indicators of brain damage or of schizophrenia, but restricted to those whose correct diagnoses are already known to be either one or the other. (16 refs.) - *B. Berman.*

Veterans Administration Hospital
St. Cloud, Minnesota

3132 **HATCH, ERIC; & FRENCH, JOSEPH L.** The revised ITPA: Its reliability and validity for use with EMRs. *Journal of School Psychology*, 9(1): 16-23, 1971.

The revised Illinois Test of Psycholinguistic Abilities (ITPA) is a fairly stable instrument for the diagnosis and remediation of psycholinguistic deficits in EMRs. In an investigation of the validity and reliability of the revised ITPA, EMRs were administered the ITPA and given it again 3 months later. Six criterion instruments were also given to explore concurrent validity. A coefficient of at least .80 was obtained for 6 of the subtests, and 4 others were above .70. The 2 subtests of visual reception and sound blending appeared to lack sufficient stability to be used diagnostically and remedially. Validity of the subtests was neither confirmed nor disproven, since in most cases there was more than 1 significant correlation between criterion tests and other ITPA subtests. This might be explained by the presence of a G factor and the heterogeneous nature of the sample. (8 refs.) - *S. Glinsky.*

Pennsylvania State University
University Park, Pennsylvania 16802

3133 **NEEMAN, RENATE L.; & PHILLIPS, HERBERT E.** Perceptual-motor attributes of mental retardates. *American Journal of Occupational Therapy*, 25(5): 253-258, 1971.

The Purdue Perceptual Motor Test demonstrated reliability and construct validity when applied to young adult MRs. By means of correlational analysis the Purdue Perceptual Motor Test was investigated for test-retest reliability and construct validity of the instrument for young adult MRs in a sheltered workshop. Test-retest reliability was adequate (76-95) for all but 3 of 20 items administered. Interrelations among items in the study were consistent with those previously obtained by Roach and Kephart sampling normal "achieving" school children. Several significant correlations among items appeared to be reflections of the frequent incidence of perceptual motor deficits in the MRs surveyed. Several items seemed to be intercorrelated with background variables of IQ, CA, or sex. (18 refs.) - *S. Glinsky.*

Cantalician Center for Learning
Buffalo, New York

3134 **EGGERT, DIETRICH.** Empirische Untersuchungen zur Psychodiagnostik der geistigen Behinderung: Entwicklund und Weiterfuhrung der Testbatterie fur geistig behinderte Kinder (TBGB) (Empirical investigations of the psychodiagnosis of mental retardation: development and continuation of the battery of tests for mentally retarded children). *Zeitschrift fur Heilpadagogik*, 9, 457-485, 1970.

The standardization of the battery of tests for MR children makes possible a reliable, differentiated performance diagnosis of MR children. Conventional IQ tests are inappropriate for MR performance diagnosis because they are designed for normal populations and do not reflect the special differences of the MR child. The 6 tests standardized were the Columbia Mental Maturity Scale, Colored Progressive Matrices, Peabody Picture Vocabulary Test, Dotting of Circles, the Following of Instructions and the Hamburger version of the Lincoln-Oseretzky Motor Development Scale. In addition, a short form of the Vineland Social Maturity Scale was utilized. Standardization was accomplished between 1965 and 1967 in the Federal Republic of Germany on 42 samples of children between 7 and 12 years of age totaling 1,209 MRs and 454 children with learning disabilities. The reliability average of the battery of tests was .93. These standardized tests produce an MR test profile in intelligence, motor performance, attention, and language which parallels closely the results of long-term remediation observation. (19 refs.) - *S. Glinsky.*

Deutsches Institut fur
Internationale Padagogische Forschung
Schlossstrasse 29-31, Federal Republic of Germany

3135 **SWARTZ, JON D.; CLELAND, CHARLES C.; DREW, CLIFFORD J.; & WITZKE, DONALD B.** The Holtzman Inkblot Technique as a measure of perceptual development in mental retardation. *Journal of Personality Assessment*, 35(4): 320-325, 1971.

The Holtzman Inkblot Technique (HIT) is a reliable measure of perceptual development in MRs. The Ss tested with the HIT were 96 MRs and normals comprising 4 criterion IQ groups ranging in IQ from 34 to 106. Each group was matched for sex and CA. The results revealed consistent monotonic changes with IQ level for 6

HIT variables, including 4 which previously had proved to be reliable and meaningful indices of perceptual development. In addition, these HIT variables confirm the nature and direction of this development. (10 refs.) - *S. Glinsky.*

University of Texas at Austin
Austin, Texas 78712

3136 **PUSTEL, G.; STERNLICHT, M.; & DeRESPINIS, M.** Tree drawings of institutionalized retardates: Seasonal and color effects. *Journal of Genetic Psychology*, 118 (Second half): 217-222, 1971.

Measurements of seasonal and color effects on tree drawings by retardates (chromatic and achromatic drawings were obtained from 240 institutionalized retardates: 60–30 adolescents and 30 adults–for each season) showed that, in every case, regardless of season, chromatic trees were drawn larger than achromatic. Color, because of its emotional implications, was more potent than season, as reflected in leaves drawn on the trees. Adult retardates drew conventional representations, regardless of season; adolescents seemed intensely alert to seasonal cycles. The unsettling problems of adolescence seem sufficiently overwhelming to counter the conforming effects of institutionalized life. Consistency of the "color effect" principle was regarded as having important psychoanalytic implications. (7 refs.) - *B. Berman.*

Willowbrook State School
Staten Island, New York

3137 **KENNY, THOMAS J.** Background interferance procedure: A means of assessing neurologic dysfunction in school-age children. *Journal of Consulting and Clinical Psychology*, 37(1): 44-46, 1971.

The Bender Visual-Motor Gestalt Test with the BIP (Background Interference Procedure) was used to assess neurologic dysfunction in 3 groups of children. Ss were 180 children (CA 8-12 years), divided into brain damaged, emotionally disturbed, and a normal control group. Each child was individually required to draw the Bender-Gestalt figures and repeat the drawings on BIP paper on which randomly intersecting wavy lines are drawn. Two psychologists were scorers, scoring by the Pascal-Suttle System as modified by Canter. Interscorer reliability was .90, based on a Pearson product-moment correlation. A difference (D) score was computed by subtracting the score of the standard administration from the BIP score. The *t* test of difference of means was used to demonstrate statistically that there was no difference between the control and the emotionally disturbed groups. The brain-injured and control groups were compared, and the *t* statistic exceeded significance at the .01 level of confidence. This result supports the major hypothesis that the *D* score for the group with cerebral dysfunction would differ significantly from the scores of the other 2 groups. The BIP is a quick and reliable way of identifying learning problems in elementary school children. The test procedure was sensitive to all variations of pathology. (5 refs.) - *T. C. Turner.*

University of Maryland Hospital
Baltimore, Maryland 21201

3138 **MILGRAM, NORMAN A.** IQ constancy in disadvantaged Negro children. *Psychological Reports*, 29(1): 319-326, 1971.

A longitudinal follow-up study with conventional intelligence scales was done on 59 disadvantaged Negro children ranging in age from 3 to 8 years in order to deal with the question of group IQ changes, the change over time for the individual child, and the relationship of IQ to the test-taking behavior of the child. When changes in IQ by individual child were computed for ages 3 to 8 and 7 to 8, the percentage of children changing by 10 or more IQ points was 40.6 for the former comparison and 20.3 for the latter ($z=2.34$, $p<.02$) in the case of the Binet. The corresponding percentages for the Peabody were 59.5 and 32.2 ($z=3.54$, $p<.001$). Test-taking behavior correlated with IQ scores obtained at the time of testing but was of little help in predicting later IQ scores. Examination of the data from this and other studies seems to indicate that IQ scores do not decline from age 3 to 8 for disadvantaged children who are not exposed specifically to enrichment programs; after age 8, a decline in IQ may be evidenced. (12 refs.) - *B. J. Grylack.*

Temple University
Philadelphia, Pennsylvania

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3139 **SALVIA, JOHN; & YSSELDYKE, JAMES.** An analysis of the reliability and validity of the Ishihara Color Plates with mentally retarded males. *Perceptual and Motor Skills*, 33(1):243-246, 1971.

Sixty-nine MR males with CA of 8 through 16 years and IQ ranging from 40 to 75 received the Ishihara Color Plates, the Peabody Picture Vocabulary Test (PPVT), and Raven Colored Progressive Matrices in an investigation of the reliability and criterion validity of the Ishihara Color Plates with boys of low intelligence. The product-moment correlation coefficient for raw score stability was .827. However, the phi coefficient for diagnostic stability was only .486. Internal consistency was .92 on the retest. Consonant with other studies using different measures of color vision with children of limited intellectual ability, very high prevalence rates were obtained with the use of the Ishihara Plates. It was demonstrated that these prevalence rates were highly inflated by the fact that large numbers of children with normal color vision failed the tests. While the Ishihara Plates showed high internal consistency and moderate to high raw score stability and validity, diagnostic stability and validity were too low for accurate screening of children of limited intellectual ability. (6 refs.) - *B. J. Grylack.*

Institute for Research on Exceptional Children
University of Illinois
Urbana, Illinois

3140 **WELLS, DONALD G.; & PEDRINI, DUILIO T.** Relationships among Wechsler Adult Intelligence Scale, Goodenough-Harris, and Peabody Picture Vocabulary tests with institutionalized retarded adults. *Perceptual and Motor Skills*, 33(1): 227-232, 1971.

One hundred and fifty MR adults ranging in age from 20 to 34 years were tested in an investigation of the relationships between the Wechsler Adult Intelligence Scale (WAIS), Goodenough-Harris, and Peabody Picture Vocabulary Test (PPVT). In the analysis of variance, the tests showed significant differences ($p<.001$), while the sex variable and the interaction between test and sex correlations (rs) showed that the PPVT IQ correlated best with the WAIS Verbal Scale IQ, while the Goodenough-Harris IQ correlated best with the WAIS Performance Scale IQ. The PPVT IQ correlated higher with the WAIS IQ than did those based on drawings. The correlations of females were generally lower than those of males when rs were computed within WAIS and Goodenough-Harris IQs and between WAIS, PPVT, and Goodenough-Harris IQs. The standard errors indicated that the PPVT IQ, a vocabulary measure, was a somewhat better predictor of the WAIS Full Scale IQ. (12 refs.) - *B. J. Grylack.*

University of Tennessee
Martin, Tennessee

3141 **COLE, SPURGEON; & HUNTER, MILDRED.** Pattern analysis of WISC scores achieved by culturally disadvantaged children. *Psychological Reports*, 29(1): 191-194, 1971.

One hundred boys and 99 girls aged 6 to 15 years, referred for psychological testing because of inability to compete in the academic area, were administered the entire Wechsler Intelligence Scale for Children (WISC) in an effort to determine which of its subtests are affected least by cultural deprivation. The scores fell into 2 groups, relatively higher scores being earned on Picture Completion, Similarities, Comprehension, and Vocabulary. Lowest scores were obtained on Information, Picture Arrangement, Math, Object Assembly, Coding, and Block Design. While the analysis of variance did not yield significant Fs, all subtests between the 2 groups did differ significantly at the .01 level. The study supported the findings of other investigators who found no differences between the verbal and performance abilities of socially and economically deprived children. There was no evidence to suggest that cultural deprivation has less detrimental effects on performance ability than on verbal or abstract skills. (13 refs.) - *B. J. Grylack.*

Clemson University
Clemson, South Carolina

3142 **VANZYL, F. J.; & IVES, L. A.** Visual perception and eyemotor coordination in a group of young deaf children. *Developmental Medicine and Child Neurology*, 13(3): 373-379, 1971.

The Frostig Developmental Test of Visual Perception is supposed to measure independent visual-perceptual and eye-motor skills and was therefore administered to 24 deaf children of mean Visual-Perceptual Quotient 80.9 in order to validate this. Tests of letter and word matching and copying were also given. The Frostig results did not differentiate children on the basis of exogenous or endogenous causes of deafness, chronological age, or sex. Factor analysis revealed only one significant variable on the Frostig and this was general perceptual maturity. Letter/word tests were found to be equally valid measures of ability and would appear to be more relevant to techniques used in teaching the deaf to read. (25 refs.) - *E. L. Rowan.*

University of Manchester
Department of Audiology
Manchester M13 9PL, England

3143 **SLEMON, A. G.** The effects of type of reinforcer, schedule of reinforcement, sex, and personality on response rates of mentally retarded children. *Journal of Mental Deficiency Research*, 15(1): 38-42, 1971.

Overconforming MR children responded at a significantly higher rate than underconforming MR children on a lever pulling task. The 12 male and 12 female Ss (age range 11 to 16.01 yrs) were divided into conforming and nonconforming groups on the basis of teacher and principal ratings. Overconforming Ss were those who were strongly influenced by teacher praise or criticism, and underconforming Ss were little influenced by these factors. Differences in reinforcement (candy or beans) or reinforcement schedules (VR 8.7; VR 17; and VR 25) did not appear to affect performance. (9 refs.) - *J. K. Wyatt.*

University of Western Ontario
London, Ontario, Canada

3144 **ALLEN, ROBERT M.; ADAMO, CYNTHIA; ALKER, LESLIE N.; & LEVINE, MARTIN N.** A study of recognition and reproduction of Bender Gestalt figures by children of average and below intelligence. *Journal of Genetic Psychology*, 119: 75-78, 1971.

A study of the ability of children of average intelligence and EMRs to recognize and reproduce the one best of 4 Bender-Gestalt model designs revealed that EMRs were inferior to normal children in both functions. Scores of EMRs on the Developmental Test of Visual Perception revealed a high incidence of visual perceptual impairment, which affected both recognition and reproduction, but reproduction more markedly. (9 refs.) - *N. Jarvis.*

Box 8341 University Branch
Coral Gables, Florida 33124

3145 **ROSEN, M.; BERGMAN, MARILYN; & TELFER, MARY A.** MMPI profiles of males with abnormal sex chromosome complements. *Journal of Mental Deficiency Research*, 15(3): 169-176, 1971.

The Minnesota Multiphasic Personality Inventory (MMPI) in card form was applied to 9 Klinefelter males and 7 XYY males for systematic personality evaluation. A control population of 50 Ss was randomly selected from a hospital for the criminally insane. The most striking findings are elevations on 2 pathological scales - Psychopathic Deviate and Schizophrenia, which occurred in both clinical groups. Klinefelter profiles were slightly more elevated than XYYs in a pathological direction, but the differences were not statistically significant. Median profiles for the 50 controls showed lower elevation peaks. From the results are concluded: (1) Median profiles of Klinefelter and XYY Ss identified in prison and hospital populations show peaks on the Pd and Sc scales of the MMPI; (2) The 2 syndromes showed much similarity in median MMPI profiles; (3) Both XXY and XYY males seem to be qualitatively similar to a control group of 50 psychiatrically disturbed prisoners; (4) median profiles of XXY and XYY males were qualitatively different from 83 non-psychotic prisoners of normal chromosome complements; (5) At present, insufficient evidence exists to conclude that a "typical" MMPI profile exists for either syndrome. Further studies must be made of XXY and XYY profiles in relation to other control groups. (31 refs.) - *T. C. Turner.*

Elwyn Institute
Elwyn, Pennsylvania 19603

3146 **ROCHE, T. F.; *SHEEHAN, P.; LYDIA; M.; WALSH, J.; & MACAIRT, J.** A study

of handicaps and their effect on lipreading among rubella-deaf girls. *Developmental Medicine and Child Neurology*, 13(4): 497-507, 1971.

Relationship between lipreading ability and intellectual functioning, visual acuity, perceptual ability, retinitis, and other sequelae of maternal rubella was investigated by giving a standardized lipreading test to 47 girls (ages 5-18 years) in a school for the hearing impaired. All Ss were born with varying degrees of deafness as a result of maternal rubella, and also had at least one other sign of the rubella syndrome. Of all the variables, only degree of rubella retinitis in the younger Ss (5-8 years) had a slight association with lipreading ability. Also studied was the relationship between the time during pregnancy at which the mother was infected with rubella and the severity of hearing loss, retinitis, and other handicaps. Two mildly significant relationships were found: the earlier the infection, the more severe the retinitis ($p=0.039$); and the earlier the infection, the less the risk of severe language disorder ($p=0.066$). (11 refs.) - *K. L. Woodworth.*

*16 Whitehorn Road
Clonskeagh, Dublin 14, Ireland

3147 **GRANTHAM-MCGREGOR, SALLY M.; & *HAWKE, W. A.** Developmental assessment of Jamaican infants. *Developmental Medicine and Child Neurology*, 13(5): 582-589, 1971.

As part of a longitudinal development survey of 300 Jamaican infants from birth onwards, 66 of the infants had full developmental assessments at 1 year of age. All developmental assessments were carried out by a pediatric neurologist and psychiatrist. Ss were evaluated by means of the Yale Developmental Schedules, and the Gesell and Armatruda general examinational procedures were employed. Infants from the higher socioeconomic group scored better than those from the lower socioeconomic group in language behavior ($p=<0.01$) and fine motor behavior ($p=<0.05$). First-born infants were found to be slightly but consistently better than later-born infants in all areas of development, the difference being significant in gross motor behavior ($p=<0.05$). The 10 heaviest male and female infants scored significantly higher in language behavior ($p=<0.01$) than the 10 lightest. Infants with a history of first spontaneous respiration within 1 month after birth were better in all areas of development than those whose initial spontaneous respiration took longer. Sex did not make any consistent difference in the performances of the infants. (16 refs.) - *B. J. Grylack.*

Hospital for Sick Children
Toronto, Canada

TREATMENT AND TRAINING ASPECTS – Educational

3148 **CARTWRIGHT, G. PHILLIP.** The relationship between sequences of instruction and mental abilities of retarded children. *American Educational Research Journal*, 8(1): 143-150, 1971.

No relationship appears to exist between sequences of instruction and the mental abilities of MR children. Forty EMR adolescents randomly assigned to 2 groups received 2 different sequences of programmed instruction. The 2 sequences, of which 1 was ordered and 1 random, consisted of 612 identical frames, but in different orders for the 2 groups. No differences were found between the 2 groups on immediate learning, retention, or transfer. Results of correlational analyses were equivocal, but there was some indication that different aptitudes were required by the 2 groups in order to make equivalent learning test scores. (10 refs.) - *S. Glinsky.*

201 Chambers Building
University Park, Pennsylvania 16802

3149 **SUNLEY, ROBERT.** Thinking skills as a goal in an after-school program. *Children*, 18(3): 90-94, 1971.

A Thinking Skills Program has been developed at the Family Community Center in Freeport, N. Y., to aid disadvantaged and culturally deprived children in developing cognitive skills (labeling, comparison, categorization, for example) which they have not yet mastered. Seven major aspects of learning theory taken into account in the program (sequence and transferability, language development, correlation of motor activity and cognitive development, activities without failure, developmental level, structure of participation, and group behavior) are described and several brief descriptions of program activities are given. The emphasis on thinking skills is applicable to many other group programs for children and requires neither special equipment nor costly materials. Groups can be led by professionals or paraprofessionals, but staff orientation and continuing supervision are necessary for programs to remain effective. (No refs.) - *T. C. Turner.*

Family Service Association
Nassau County, New York

3150 **SPICKER, HOWARD H.** Intellectual development through early childhood education. *Exceptional Children*, 37(9): 629-640, 1971.

Examination of the effects of 4 preschool curriculum models (traditional, cognitive development, perceptual motor development, and academic skills) aimed at increasing intellectual development and school achievement revealed that programs based on cognitive or academic skill development produced the largest increase in IQ scores among disadvantaged children. When traditional curriculum programs had specific short- and long-term goals, included language development, and were highly structured and well-supervised, significant intellectual growth occurred. Structured programs which were not cognitively or academically oriented resulted in intellectual gains only when there was a strong emphasis on oral language development. Preschool academic skill oriented programs which produced rote reading and arithmetic computation skills did not result in improved reading comprehension or arithmetic reasoning skills. Success in the primary grades of many inner city ghetto and rural community schools appears to depend on the development of fine motor, memory, and general language abilities rather than on abstract reasoning, critical thinking, or creative thinking. Other variables which contribute to intellectual development included home intervention programs, large proportion of adults to children in preschool programs, and increased periods of preschool intervention. (16 refs.) - *J. K. Wyatt.*

Indiana University
Bloomington, Indiana

3151 **OLSHIN, GEORGE S.** Model centers for preschool handicapped children — Year II. *Exceptional Children*, 37(9): 665-669, 1971.

The purpose of the 43 Model Centers Programs funded during the first 2 years after passage of the Handicapped Children's Early Education Assistance Act of 1968 is to develop a variety of models to demonstrate comprehensive quality services to young handicapped children (CA 0 to 8 yrs) and their families. Some centers provide identification and screening services. A wide variety of instructional and remedial approaches is used, including prescriptive teaching, behavior modification, and diagnostic evaluation. The centers train and use paraprofessional aides and provide training and counseling for parents. They also conduct personnel training programs. Project evaluation is one of the most difficult tasks. Standard and supplementary assessment instruments are in use. (3 refs.) - *J. K. Wyatt.*

Southern Connecticut State College
New Haven, Connecticut

3152 **BLUM, EVELYN R.** Conversations in training and research. Interview with George Sheperd: An R&D Center focuses on early childhood education. *Exceptional Children*, 37(9): 671, 676-680, 1971.

The program of the Early Childhood Research and Development Center at the University of Oregon is directed toward very young handicapped preschool children and is the only center in the National Program of Early Childhood Education that has assumed handicapped children as a primary responsibility. Program focus is on the identification, modification, and prevention of handicapping conditions that preclude or interfere with successful academic performance. Programs include children from birth until age 8 or 9, although the present target population is ages 2 to 9 years. Among the ongoing research projects are studies to identify and measure observable achievement-related behaviors in grades 1 through 3; to develop intervention programs for children with deviant classroom behavior; to develop teaching programs for parents of preschool multiply handicapped children; and to find alternatives to traditional disability grouping systems. (No refs.) - *J. K. Wyatt.*

University of Oregon
Eugene, Oregon

3153 **McDONALD, PHYLLIS L.; & SOEFFING, MARYLANE.** Prevention of learning problems: Capsule summaries of research studies in early childhood education. *Exceptional Children*, 37(9): 681-686, 1971.

Eight ongoing research projects were designed to investigate the effects of early environment on later academic achievement. Among the areas under investigation are the effects of a supplemental linguistic environment based on distancing experiences on the cognitive development of disadvantaged 2-year-olds; the problem-solving styles of 4- to 8-year old children; the relationship between early social development and later academic performance; and the effects of the enriched academic environment provided by a Follow-Through Program on disadvantaged children. Other programs are concerned with the early identification and remedial treatment of developmental disabilities; professional and paraprofessional training; altering the home environment through a parent-child development program; and the effects of a planned variation in a Head Start Program. (No refs.) - *J. K. Wyatt.*

No address

3154 **BAER, DONALD M.; & GUESS, DOUG.** Receptive training of adjectival inflections in mental retardates. *Journal of Applied Behavior Analysis*, 4(2): 129-139, 1971.

A technique of differential reinforcement was administered to 3 institutionalized SMR children in an attempt to teach them to identify correctly the quantitative relationships between stimuli, as indexed by comparative and superlative adjectives. The study tested the concept that successful instruction in this area could result in an organized set of responses that the child then could apply correctly to new stimuli, thereby generating response to new words within his old grammar without further training. Training materials consisted of 19 sets of pictures; each set contained 4 cards which displayed quantitatively different pictures of the same basic stimulus. In general, a decreasing number of trials was required for each S to reach each successive descriptive criterion, with the minimum number being achieved repeatedly by the end of training.

It was found significant that their learning generalized to stimulus combinations or transpositions which had not been trained directly but which exemplified the same dimensions that characterized the original training experiences. (4 refs.) - *B. J. Grylack.*

University of Kansas
Lawrence, Kansas 66044

3155 **RYDBERG, SVEN.** Beginning reading discrimination taught at IQ 35 by conditioning. *Perceptual and Motor Skills,* 32(1): 163-166, 1971.

Six SMR children were able to learn complex visual discriminations fundamental to reading by simple conditioning with cards. They were initially rewarded for pairing the stimulus word with its picture. Learning was not evident until after 7 days of the 17 session program, but there was good retention in the 3 best students 24 and 48 days later. These children had MA 3.5 to 4.0, which appeared to be the major variable in their ability to learn. (1 ref.) - *E. L. Rowan.*

University of Stockholm
Stockholm, Sweden

3156 **PETER, LAWRENCE J.; & WISELY, FORREST G.** The prescriptive teaching system: A teacher education program. *Focus on Exceptional Children,* 3(3): 1-8, 1971.

A pioneer effort in applying systems analysis to teacher education resulted in the Prescriptive Teaching System. It consists of 3 phases: Individual, classroom, and therapeutic instruction. The graduate of these phases is competent to tutor an individual child, conduct classroom learning, and instruct and counsel handicapped pupils and their parents. The system is provided with built-in accountability where each component is analyzed and evaluated to determine its effectiveness in moving the system toward specified objectives. Several research programs which investigated the outcomes of the PTS are described and the need for further research is noted. (12 refs.) - *T. C. Turner.*

John Tracy Clinic
Los Angeles, California

3157 **SCAGLIOTTA, EDWARD G.** Developing the process of thought with the learning disabled child. *Focus on Exceptional Children,* 3(3): 9-10, 1971.

Simultaneous sense stimulation is suggested as a way to formulate an objective or purpose in order to initiate the process of thought in a child. In presenting ideas, the simple must precede the complex in successive steps, with the significant parts becoming significant wholes. Conditioning is also essential for the learner's development. Modeling can also stimulate a child to correct response. The child must be involved in his own learning goals so that he will have continuous positive response to the learning environment. Teacher understanding of a child's emotions during times of learning problems is necessary to maintaining of clear, logical thought. A learning disabled child can make gains in the thinking and learning process when the teacher works with his individual characteristics towards achieving functional success. (8 refs.) - *T. C. Turner.*

The Midland School
North Branch, New Jersey

3158 **SHEPERD, GEORGE.** The early education of handicapped children. *Focus on Exceptional Children,* 3(1): 1-9, 1971.

The positive effect early education has on the total development of the handicapped child is noted. Included in the discussion are: the rationale for early education; federal funding of special education programs; currently operative early education research centers and their objectives, strategies, and findings. The necessity of family involvement in the learning process is also stressed, and 10 recommendations of leading educators in the field of early education are enumerated. (40 refs.) - *T. C. Turner.*

University of Oregon
Center for Research and Demonstration in the Early Education of Handicapped Children
Eugene, Oregon

3159 **SIDMAN, MURRAY.** Reading and auditory-visual equivalences. *Journal of Speech and Hearing Research,* 14(1): 5-13, 1971.

An institutionalized 17-year-old microcephalic and SMR male participated in an experiment designed to demonstrate that certain learned auditory-visual equivalences are sufficient prerequisites for the emergence of reading comprehension, even without explicit teaching of reading comprehension. After being taught to match spoken and printed words, the reading comprehension and oral reading ability of the S improved greatly. He was then able to match picture samples to printed word choices, to match printed word samples to picture choices, and to name printed words without additional teaching. The identification of sufficient prerequisites for reading comprehension suggests that both auditory-visual equivalences can be taught completely without the intervention of a teacher. (9 refs.) - *B. J. Grylack.*

Massachusetts General Hospital
Boston, Massachusetts

3160 **GOYEN, JUDITH D.; & LYLE, J. G.** Effect of incentives and age on the visual recognition of retarded readers. *Journal of Experimental Child Psychology*, 11(2): 266-273, 1971.

A tachistoscopic form recognition task was administered to 24 retarded readers (experimentals) and 24 normal readers (controls) to examine the effect of motivation and distractibility upon the performance of these readers on a visual recognition task. Within each group, half of the Ss were aged 7.25 to 8.25 years, and half were aged 8.5 to 9.5 years. Half of the Ss from each group were tested under incentive treatments, and half under nonincentive conditions. Analysis of variance of recognition scores indicated that experimentals performed significantly less well than controls, with younger Ss performing significantly less well than older Ss. Latency of response was found to be unrelated to incentives and the other experimental variables. An overall practice effect ($p<.01$) was noted, but it had no differential effect upon the groups. The finding that retarded readers were not characterized by a greater proportion of reversal errors than normal Ss is probably due to the elimination of labeling and motor components from this visual recognition task. (13 refs.) - *B. J. Grylack.*

Macquarie University
New South Wales, Australia

3161 **GOYEN, JUDITH D.; & LYLE, J. G.** Effect of incentives upon retarded and normal readers on a visual-associate learning task. *Journal of Experimental Child Psychology*, 11(2): 274-280, 1971.

A visual-visual paired associate learning task based on 6 trials was administered to 28 retarded readers and 28 normal readers aged between 6 and 7 years, with half of the Ss from each group tested under a nonincentive treatment, and half under an incentive treatment. A highly significant overall difference between incentive and nonincentive treatments in favor of the former and a highly significant difference in learning rate over the 6 trials in favor of the incentive treatment were found. There was no significant overall difference between retarded and normal readers in ability to learn the visual associate task. The results indicate a lack of significant difference in learning rate and of significant differential response to incentives among the 2 groups. (11 refs.) - *B. J. Grylack.*

Macquarie University
School of Education
New South Wales, Australia

3162 **HARING, NORRIS G.** The systematic use of contingencies in classes for children with academic and social response deficits. *Educational Technology*, 11(4): 54-58, 1971.

Contingency management techniques were successful in classes for children with academic and social response deficits. Experimental programs were conducted with both MR and regular classes using base line measurement, continuous recording, and contingency management with token reinforcement. Programmed texts and teacher-made programs formed the basis of instruction. The results indicated that the proper use of contingency management procedures can shape appropriate academic and social classroom behavior. (8 refs.) - *S. Glinsky.*

University of Washington
Seattle, Washington

3163 **ANDERSON, RICHARD D.** Application of educational rhythmics to therapeutic recreation service. *Therapeutic Recreation Journal*, 5(2): 75-78, 1971.

Techniques of educational rhythmics, offering a positive and self-reinforcing means of reaching reluctant and retarded learners, are discussed. Since a child must first be reached before he can be taught, the language of movement, or rhythmics, can be used to reach the MR child. The experiences and movements of MR or physically handicapped children can be coordinated with simple exercises done to a background of music. Movement can be used as an effective aid to learning experiences and to making them more enjoyable for MR or handicapped children. (11 refs.) - *T. C. Turner.*

Hubbard Child Development Center
Anchorage, Alaska

3164 **HERB, MARILYN CAMP; & BRANNAN, STEVE A.** Incorporating recreation into teacher training programs: Reflections after a pilot program. *Therapeutic Recreation Journal*, 5(2): 57-59, 1971.

An 8-week summer program developed to certify and train teachers in special education incorporated recreation skills into an education program and established the importance of the recreation-education concept in the development and training of EMR children. The program is described, values and limitations are discussed, and recommendations made for future implementation. (No refs.) - *T. C. Turner.*

Oregon State Hospital
Salem, Oregon

3165 **LILLY, M. STEPHEN.** A training based model for special education. *Exceptional Children*, 37(10): 745-749, 1971.

An alternative is set forth to the special services model presently used by special education in working with EMR, emotionally disturbed, behaviorally disordered, brain injured, or learning disabled children who have been referred from regular education programs because of behavior or learning problems noticed by the teacher. The function of the new model would be to provide training for classroom teachers, who would be aided by instructional specialists in learning how to deal with problems in the class, rather than removing a problem child from the regular classroom situation for individual work. The instructional specialist, employed by and based in the school, would replace the special education teacher. (1 ref.) - *T. C. Turner.*

University of Oregon
Eugene, Oregon

3166 **BARNGROVER, ELAINE.** A study of educators' preference in special education programs. *Exceptional Children*, 37(10): 754-755, 1971.

Fifty educators were interviewed in March, 1970, to determine whether they thought that special classes for the mildly exceptional child are adequate or that he would fare better by being in a regular classroom. Twenty-seven of 50 thought the present special classes program should be retained. Reasons cited for this were: fewer frustrations for the child, less upset in the regular classroom, more specialized attention, and individualized help. More materials, qualified teachers, and smaller classes were improvements suggested. Reasons given by the 23 who supported placement in a regular school were: widened horizons and greater stimulation for the child and more good behavior models to imitate from peers. Improvements suggested included team teaching, smaller classes, and flexibility in grading and scheduling. Significant differences (chi square=6.40, df=1, p=.05 level) were found between views of teaching and nonteaching personnel. The former favored retention of special classes while the latter did not; however, reasons for this are not apparent in the study. (No refs.) - *T. C. Turner.*

Clermont County Board of Education
Batavia, Ohio

3167 **CARTER, JOHN L.; & MILLER, PHILLIS K.** Creative art for minimally brain-injured children. *Academic Therapy*, 6(3): 245-252, 1971.

In June 1968, a special class of minimally brain-injured (MBI) children was formed at the University of Houston Clinical Education Laboratory to discover what might be the effects of a creative art program on MBI children and to test the theory that such a program could improve perceptual-motor coordination. It was thought that, since the development of perceptual awareness and manipulative skills is encouraged by art and creative activities, Ss who participated in

them might improve themselves in these perceptual motor areas. During the 5-week session, 4 series of assignments were given to encourage the children to experience their world through all senses and translate this experience into drawings. Significant gains were shown by comparing pretest and posttest scores on the Marianne Frostig Developmental Test of Visual Perception. However, there is not yet a test to measure the intangible qualities of joy of learning and desire to participate in learning activities which affected the Ss in the art class. As a way of improving visual - perceptual abilities, it would seem that creative art activities are very successful, because motivation to learn comes from the pleasure of doing the task in art, and reinforcement in the form of outside rewards or teacher approval is not necessary. (3 refs.) - *T. C. Turner.*

University of Houston
Hosuton, Texas

3168 **MacMILLAN, DONALD L.** The problem of motivation in the education of the mentally retarded. *Exceptional Children*, 37(8): 579-586, 1971.

One reason why special classes for educable mentally retarded (EMR) children have failed to stimulate substantial academic or social growth may be that the contribution of motivation to performance and learning in EMRs has been overlooked. Three motivational variables are discussed (expectancy for failure, positive and negative reaction tendencies, and outer-directedness), and the findings are presented. The failure of the self-contained special class to educate the EMR child may lie partly in the lack of balanced emphasis on cognition and motivation, by treating all mentally retarded alike and ignoring individual differences. Since the EMR child experiences an unusual amount of failure prior to placement in a special class, the classroom must be adapted to allow the child to recapture his self-worth and establish a series of "success experiences" in completing given tasks. (35 refs.) - *T. C. Turner.*

University of California, Riverside
Riverside, California

3169 **SCHWARTZ, LOUIS.** A clinical teacher model for interrelated areas of special education. *Exceptional Children*, 37(8): 565-571, 1971.

The model described for the preparation of clinical teachers for interrelated areas includes behaviorally defined and measurable instructional objectives and provides behaviorally defined specifications of desired competencies, criterion measures for assessing entry and proficiency, and instructional options available for achieving these objectives. A program of studies has been constructed from the junior undergraduate through the doctoral level. Evaluation of the model will be on 2 levels: how effective the training program is in aiding the students to attain goals set forth by the model, and how effective the teacher is in helping pupils achieve the curriculum and social goals specified for them. (31 refs.) - *T. C. Turner.*

Florida State University
Tallahassee, Florida

3170 **ETIENNE, JERALD; & MORLOCK, DAVID A.** A pre-vocational program for institutionalized mental retardates. *American Institute for Mental Studies Training School Bulletin*, 67(4): 228-234, 1971.

A comprehensive prevocational program at Dixon State School for MRs in Dixon, Illinois, considers each student's integrities and deficiencies in facilitating his return to the community. Starting with the notion the student knows little of the outside world, the program takes him in sequential phases through a series of vocational and living situations that assure maximum success at each level until eventual discharge from the institution. The progression goes from the sheltered life in the general population cottages to the halfway houses to community placement. Program goals include development of self-concept, personal attitudes and community adjustment, and skills, including dexterity, coordination, task performance, counting, and acceptance of criticism. The program uses interdisciplinary skills of educators, psychologists, rehabilitation counselors, social workers, and house parents. (No refs.) - *B. Berman.*

Dixon State School
Dixon, Illinois

3171 **MEYEN, EDWARD L.; FRANK, ALAN R.; & RODEE, MYRON.** Special class teachers as field testing evaluators. *American Institute for Mental Studies Training School Bulletin*, 67(4): 198-205, 1971.

Nineteen special-education teachers successfully used instructional packages designed to teach junior-high-age MRs about law and authority (specifically shoplifting) and demonstrated the willingness of teachers to participate in such evaluations and their effectiveness in communication. The study, conducted by the University of Iowa's Special Education Curriculum Development Center, was a response to the rapid growth in special-education programs for exceptional children. The teachers, who used 3 procedures in reporting evaluations (a general evaluation report, a lesson-plan evaluation card, and written comments), showed remarkable agreement on the strengths and weaknesses of the instructional material. Pre- and post-tests showed significant improvement in pupils' knowledge of law and authority. (4 refs.) - *B. Berman.*

University of Missouri
Columbia, Missouri

3172 **KOKASKA, SHAREN METZ; & KOKASKA, CHARLES J.** Individualized work centers: An approach for the elementary retarded child. *Education and Training of the Mentally Retarded*, 6(1): 25-27, 1971.

The concept of classroom interest centers was adapted for use with primary EMR students. In a class of 10 students (6 1/2-11 years, IQ 50-75) who were all functioning at a readiness or first grade level, 5 work centers were arranged about a central area for desks. Each center contained a certain type of task: perceptual tasks, taped exercises in auditory discrimination, storybook records and followup work to increase listening skills, manipulative tasks, and art activities. Each student was assigned to a different center each day of the week, for 1/2 hour. Reasons for using this approach were to divide students into smaller groups to facilitate instruction, to help students become more reliant on their own initiative, and to provide special training. A teacher's aide was used so that reading and arithmetic could be taught to small groups while other students were at the work centers. A reward system (checkmarks traded for a prize) was used at first to reinforce attendance at the work center, but after 3 months it was no longer necessary. The learning center approach was found to be a useful technique in approaching the ideal of prescriptive teaching. (1 ref.) - *K. L. Woodworth.*

Palos Verdes Unified School District
Palos Verdes Peninsula, California

3173 **DEZELLE, WATER.** The credibility of programmed material as an instructional technique with mentally retarded pupils. *Education and Training of the Mentally Retarded*, 6(1): 16-19, 1971.

The feasibility of using programmed instruction in mathematics as a teaching technique for EMR junior high school students was examined. Ss were EMR students in 5 classes from 5 different school districts; students in control classes were matched for age, sex, race, IQ, and pretest scores on the California Arithmetic Test (CAT). Programmed instruction was used in the experimental classes for 4 1/2 months, while a variety of conventional teaching techniques was used in the control classes. Ss in experimental classes adjusted to the format and curriculum of the programmed materials and achieved a level of self direction and self discipline adequate to the demands of the programmed materials. Post-testing with an alternate form of the CAT revealed no significant differences in the gains of the two groups, but programmed instruction was found to qualify as a feasible instructional technique for this population. (9 refs.) - *K. L. Woodworth.*

Lamar State College of Technology
Beaumont, Texas

3174 **MONROE, J. DONALD; & HOWE, CLIFFORD E.** The effects of integration and social class on the acceptance of retarded adolescents. *Education and Training of the Mentally Retarded*, 6(1): 20-24, 1971.

Two variables related to the social acceptance of EMR students in an integrated junior high school were investigated: length of time of integration and social class. Ss were 70 EMR boys (13-16 years, IQ 54-92) in 19 physical education classes (with a total of 574 unretarded students) in 5 different junior high schools with integrated programs. The Ohio Social Acceptance Scale (OSAS) was administered by the physical education teachers, and the Socioeconomic Index for Occupations (SIO) was used to estimate social class. Results showed that the social acceptance of the retarded Ss was the same whether the Ss

had been integrated for 1,2, or 3 years. However, social class was positively related to social acceptance, since the difference between the mean OSAS scores for the high (SIO score>16) and low mean (SIO score<16) socioeconomic EMR groups was significant. (14 refs.) - *K. L. Woodworth.*

Oshkosh Area Public Schools
Oshkosh, Wisconsin

3175 **BROWN, LOU; & FOSHEE, JAMES G.** Comparative techniques for increasing attending behavior of retarded students. *Education and Training of the Mentally Retarded*, 6(1): 4-11, 1971.

The acquisition of attending behavior regardless of the properties of the stimuli was tested with retarded children in the classroom. The questions investigated were whether attending behavior of Ss toward peripheral classroom stimuli (pictures on a wall) could be increased without manipulating the physical properties of the stimuli, and whether individual reinforcement was more effective than reinforcement of a model in the presence of observing classmates. Three classes of 7 Ss each (age 7-14 years) were randomly assigned to 3 conditions: individual training, model training, and control (in which the stimuli were merely exhibited). During each of 1 baseline week, 3 training weeks, and 1 retention week, Ss were asked to recognize the one correct picture among 4 pictures after the one picture had been removed from exhibition in their classroom. Ss in the individual and model training conditions performed significantly better ($p<.05$) than the controls, but no significant differences in attending were found between Ss receiving individual and model training. Since model training can be as effective as individual training in certain circumstances, it is suggested for consideration as an economical instructional technique. (23 refs.) - *K. L. Woodworth.*

University of Wisconsin
Madison, Wisconsin

3176 **JONES, REGINALD L.** Preferences for teaching intellectually exceptional children. *Education and Training of the Mentally Retarded*, 6(1): 43-48, 1971.

Factor analytic techniques were used to determine relationships between preferences for teaching EMR children and for teaching certain grades or subjects. Ss were 141 practicing EMR and regular class teachers and 399 undergraduate regular education students. Ss completed a ranking form, indicating the groups or subjects they most preferred to teach. The factor Preference for Teaching EMR, found in all samples, included high interrelationships among preferences for elementary, junior high, and senior high teaching of the retarded. A bipolar factor comprised the elementary and secondary level specialties, with one exception: interest in teaching secondary age EMRs was not related to general secondary teaching interest. That preferences for teaching elementary EMRs were, however, related to general kindergarten-elementary teaching preferences suggests that potential teachers of the elementary level EMR may be most easily recruited from among already employed elementary school teachers. (16 refs.) - *K. L. Woodworth.*

University of California
Riverside, California

3177 **BLUM, EVELYN R.** The Madison Plan as an alternative to special class placement: An interview with Frank Hewett. *Education and Training of the Mentally Retarded*, 6(1): 29-42, 1971.

The Madison Plan, now operating in the Santa Monica (California) Public Schools, extends some of the ideas of the engineered classroom to the field of special education. The plan provides an administrative and instructional framework within which students with various handicaps (mostly EMR and educationally handicapped) can be combined into a single program. Rather than being labeled as to handicap, students are classified along the dimension of readiness for regular class functioning: Preacademic 1, Preacademic 2, Academic 1, Academic 2. Beginning with a self contained engineered-type classroom with high levels of reinforcement at Preacademic 1 level, students progress along the continuum as they are able, with total integration into a regular classroom (Academic 2) the ultimate goal. With 2 teachers and 2 aides, the program can handle about 36 students. Emphasis is placed on obtaining the cooperation and assistance of the regular classroom teachers, who may have the EMRs in their classes for at least an hour each day. The program has been successful in integrating the special students into regular classes for

greater lengths of time. (10 refs.) - *K. L. Woodworth.*

CEC Information Center
Arlington, Virginia 22202

3178 **U.S. Health, Education, and Welfare Department.** *Categories of Exceptionality and Disorders of Learning and Behavior Found in Educational Follow-up Population.* Rubin, Rosalyn; Balow, Bruce; & Wasson, John. Office of Education, Bureau of Research, 1970. 10 p.

A survey of the incidence of specified learning and behavior problems in 967 children (their mothers received prenatal care and were delivered at the University of Minnesota Hospitals) covered by the Educational Follow-up Study which reported the association between perinatal variables and later occurrence of school learning and behavior problems, showed scores closely approximating those of normative samples. Almost 44 percent of Ss displayed one or more of these elements: a steady increase in special-class placements, repetition of at least one grade, and manifestation of attitude and behavior problems (boys consistently showed more problems than girls). Stanford Paragraph Meaning scores followed quite closely score distributions for normative samples. Ss scoring one or more years below actual grade placement were identified as reading disability cases. The problems uncovered suggested a fertile source for study of learning and behaviour problems, and stimulated questions concerning base rates for such problems in the general population. (No refs.) - *B. Berman.*

University of Minnesota
Minneapolis, Minnesota

3179 **GAY, WILLIAM O.; & STEPHENSON, BOBBY L.** A systems approach as a method of training teachers of mentally retarded children. *Education and Training of the Mentally Retarded,* 6(2): 56-66, 1971.

Evaluation of research on the merits of placing MR children in special or regular classes has indicated that it is the instruction that these children receive in the classroom which determines their social and academic development. Some colleges, in their preparation of teachers, have developed curricula that incorporate certain aspects of instructional objectives. The Outcomes Charts approach is based upon stated competencies in arithmetic and social, communication, safety, health, and vocational areas for preschool, primary, intermediate, prevocational, and vocational classes for MRs. A very promising approach towards solving the learning problems of the retarded is the application of the systems methodology. The 4-phase system which uses a taxonomy of learning tasks is based upon a transition from idea to objective to classification of learning type, and finally to an evaluation of output competencies. This approach incorporates systematic use of the body of empirical knowledge of learning in order to maximize the probability that instructional procedures will be selected which are efficient in promoting attainment of the desired instructional objectives. The system also includes features designed to prevent preconceived assumptions about the background knowledge of the pupil, and it leads to a focus upon the individual pupil. (10 refs.) - *B. J. Grylack.*

Northeast Louisiana University
Monroe, Louisiana

3180 **CHRISTIANSEN, TED.** An integrated undergraduate program in mental retardation. *Education and Training of the Mentally Retarded,* 6(2): 67-69, 1971.

The teacher preparation program in MR at Madison College, Harrisonburg, Virginia, is a 4-year undergraduate program designed to train students to teach elementary school EMR pupils through a series of field experiences combined with class work. Students take an introductory course in special education in their freshman year which serves as a screening device in the program. Students enroll for 2 special education courses in the sophomore year and an additional 3 in the junior year together with special education electives. The 3 integrated lecture-field experience courses in the senior year culminate in student teaching. (No refs.) - *B. J. Grylack.*

Madison College
Harrisonburg, Virginia

3181 **HRUBI, JOSEPH R.** Work stations for job-related skills. *Education and Training of the Mentally Retarded,* 6(2): 70-73, 1971.

Sixty-one EMR students are currently enrolled in the Somerville (Massachusetts) Occupational Opportunity School, a public school designed to train MR youth for suitable occupations and then to place them in business, industrial, and government positions. Two special educators are involved in the instruction program of related occupational education, which emphasizes in particular the psychosocial and sociophysical development of the individual and his personal adjustment training. Instruction is provided in the areas of food service, transporting and storing, woodworking and furniture refinishing, home mechanics, and garment production and alteration. Funds obtained for the program from the Bureau of Special Education of the Massachusetts Department of Education and the Federal Office of Social and Rehabilitation Service have been used to expand the services and facilities at the occupational school. (No refs.) - *B. J. Grylack.*

Somerville Public Schools
Somerville, Massachusetts

3182 **BROWN, LOU; & PERLMUTTER, LUCILLE.** Teaching functional reading to trainable level retarded students. *Education and Training of the Mentally Retarded,* 6(2): 74-84, 1971.

Seven TMR students ranging in age from 14 to 19 (X=16), in IQ scores from 39 to 51 (X=44), and in years in school from 2 to 12 (X=9.2) participated in a functional reading demonstration in which they were presented with a series of printed stimuli (sentences) and were requested to label these stimuli and to do what the sentences indicated. The first part of the program determined how well each S could label the words in sentence order, respond differentially to the sentence after labeling the words, and respond differentially to the sentences after listening to the teacher label the words. For the second and third parts of the program, the 7 Ss were assigned randomly to 2 groups and were taught to label the words correctly in the order in which they occurred in the sentences and, finally, to respond differentially to each of 9 sentences. The 2 groups attained a 43 and 55% listening vocabulary efficiency level, respectively, for the first 3 trials on this task but responded perfectly during the following 3 trials. It appears that reinforcement principles are a valid and relevant source of instructional technology and that the systematic utilization of these principles with groups of children in public school classrooms can result in acquisition by TMR students of academic skills that had been excluded previously from curricula. (9 refs.) - *B. J. Grylack.*

University of Wisconsin
Madison, Wisconsin

3183 **JORDAN, JUNE B.** Current and significant: The Curriculum R & D Center in Mental Retardation. *Education and Training of the Mentally Retarded,* 6(2):85-87, 1971.

The main purpose of work carried out at the Curriculum Research and Development Center in MR, Yeshiva University, is to improve educational provisions and programs for MR children in special classes, regular classes, and in tutorial settings. Currently, its central goal is to develop a comprehensive, developmentally organized educational program for EMRs to be used from preschool through secondary programs. In addition to *Centerline,* the newsletter of the Research and Development Center, other important publications on the education of gifted and handicapped children include *Exceptional Child Education Abstracts, Teaching Exceptional Children,* and *Dimensions,* published by the Council for Exceptional Children. (No. refs.) - *B. J. Grylack*

Yeshiva University
New York, New York 10003

3184 **HEISS, WARREN E.; & MISCHIO, GEORGE S.** Designing curriculum for the educable mentally retarded. *Focus on Exceptional Children,* 3(2): 1-10, 1971.

The results of studies undertaken to determine the efficacy of special class placement for EMR children have constituted an influential factor in the growing concern to provide meaningful curriculum materials for these children. The ultimate goal is to educate EMR children to perform adequately in several social learning contexts. Performance in those contexts is a reflection of problem-solving skills attained in various curriculum activity areas. The mastery of problem-solving skills is a function of the acquisition or use of basic psychoeducational processes. In practice, the classroom teacher can observe performance in a social behavior context, can decide which curriculum activity area is contributing to inappropriate performance, and, if necessary, can

analyze the curriculum activity area in terms of influential psychoeducational processes. A fully integrated social learning and skills program provides the contextual interface whereby researchers can assess student processing, curriculum developers can evaluate the flow of concepts and skills, and teachers can manage learning through a systematic, total educational program. (22 refs.) - *B. J. Grylack.*

Yeshiva University
New York, New York

3185 **CONNOLLY, AUSTIN J., ed.** Classroom forum. *Focus on Exceptional Children*, 3(2): 12, 1971.

Interaction, involvement, flexibility, and creativity can provide the material by which a teacher of an EMR special class can help to minimize the special class stigma. If the special class goes on a field trip, other students should be invited if there are extra spaces. A tutorship program can be developed using students from the upper grades, particularly the hyperactive students, as tutors. The special class teacher should become an integral part of the faculty groups. If the special classroom is equipped with particular items of interest to other students, they should be invited to do project work with the special class. It is the action undertaken by the teacher which ultimately determines whether his or her special class will be accepted by other children. (No refs.) - *B. J. Grylack.*

University of Missouri
Columbia, Missouri

3186 **BOWLEY, AGATHA H.; & GARDNER, LESLIE.** *The Handicapped Child.* (Third edition) Edinburgh and London, Churchill Livingstone, 1972, 203p.

Utilizing primary experience and literature sources, this volume, written by psychologists working in association with medical and teaching personnel, offers findings on 6 groups of handicapped children, and is presented in an effort to allay anxieties usually surrounding such children. The approach focuses on compassion and love in caring for the handicapped, and in training informed adults to provide necessary understanding and skill for such children, whose dependency needs are large and who, nevertheless, have a right to develop their abilities. This third edition enlarges the coverage of its predecessor, but retains its emphasis on cerebral palsy because of its complexity and recent therapeutic and educational developments. (185 refs.) - *B. Berman.*

CONTENTS: Cerebral Palsy; Brain Damage; Spina Bifida; Deaf Child; Blind Child; Autistic Child; Handicapped Child in Family and Community.

3187 **LIVESEY, P. J. & LITTLE, AUDREY.** Sequential learning by children. *Journal of Genetic Psychology*, 118 (First half): 33-38, 1971.

Characteristics of sequential learning observed in Rhesus monkeys was compared with observations of 2 groups of kindergarten children using the Wisconsin General Test Apparatus. The criterion of success was the same for children as for monkeys. A group was trained on a 4-response double alternation task and another on a comparable single alternation task, which, unlike in monkeys, was mastered more readily than the former task. On attainment of criterion, children were given extended sequences of 8 and then 12 responses in length. Learning one sequence, such as single alternation, seemes to facilitate—as with cats and monkeys—learning the other sequence (double alternation). Verbalization by children indicated that they established a verbal representation of the sequence. (9 refs.) - *B. Berman.*

University of Western Australia
Nedlands, Australia

3188 **MORLOCK, D.; & TOVAR, C.** Sex education for the multiple handicapped as it applies to the classroom teachers. *American Institute of Mental Studies Training School Bulletin*, 68(2): 87-96, 1971.

Sex education for the multiply handicapped and MR is considered. It should be an approach directed to their level of ability and sensitive to the environment in which they live. Four content areas should be considered as basic curriculum: awareness of self, physical changes and understanding of self, peer relationship, and responsibility to society. The instructor must be comfortable with this area of the curriculum. Sex

education should be offered in accordance with the age and intellectual capacity of the individuals and the need which they may have. Answers given to a child asking questions should be as simple as possible. In imparting sex education to adolescents, it is preferable to separate the sexes. The main purpose of sex education at this time is to minimize the development of anxiety of pathological proportions. This education should embrace the total interaction of the adolescent with his environment. Some problems in learning are: decrease of attention span; difficulties in socializing with people, in learning, and in the ability to apply knowledge to useful purpose; and difficulty in accepting new situations. For the severely handicapped child, sex education can be done only at play therapy sessions, and the process must be repeated numerous times. Cases of multiply handicapped, SMR, and PMR individuals with problems about sexuality demand the intervention of a psychiatrist. A list of 21 pamphlets concerning sex education is included at the end of the article. (44 refs.) - *T. C. Turner.*

Dixon State School
Dixon, Illinois

3189 **GESCHWIND, NORMAN.** Dyslexia. *Science*, 173(3993): 190, 1971. (Letter)

Comments are made in support of the research reported by Rozin, Poritsky, and Sotsky (March 26, 1971, *Science*) entitled "American children with reading problems can easily learn to read English represented by Chinese characters." Dyslexia is uncommon in Japan, possibly because the Japanese use an ideographic script (kanji). Japanese dyslexic children have greater difficulty learning the kanji than the kana (syllabary script), also supporting the thesis that blending of phonetic elements presents special difficulties for the dyslexic. There is also evidence suggesting that the processes of handling the complex visual patterns of ideographs and of blending smaller phonetic elements are handled differently by the nervous system. Half a century ago Bishop Harmon (quoted in Hinshelwood's 1970 monograph on childhood dyslexia) suggested that children with this problem should be taught on the plan of the Chinese. A half century later, his plan has been carried out literally. (No refs.) - *T. C. Turner.*

Boston City Hospital
Boston, Massachusetts 02118

3190 **BEREL, MARIANNE; DILLER, LEONARD; & ORGEL, MARILYN.** Music as a facilitator for visual motor sequencing tasks in children with cerebral palsy. *Developmental Medicine and Child Neurology*, 13(3): 335-342, 1971.

Thirty-four children with cerebral palsy (mean IQ 66) were instructed to repeat patterns on a xylophone (8-note diatonic step ladder). This was presented with and without sound in 4 different positions. There was significantly better performance with auditory feedback for all children. The more intelligent children also performed better on a muffled instrument. Previous studies of multimedia stimuli have shown difficulty in audio-visual matching but the concept of an auditory stimulus as a facilitator of visual learning, especially in MR subjects, bears further study. (13 refs.) - *E. L. Rowan.*

United Cerebral Palsy of New York, Inc.
1555 Linden Blvd.
Brooklyn, New York 11212

3191 **ALBERMAN, EVA D.; & GOLDSTEIN, H.** The early recognition of handicapping disorders in childhood. *Developmental Medicine and Child Neurology*, 13(3): 396, 1971. (Letter)

Two separate points are frequently confused in discussing "at risk" registers of potentially handicapped children. Efficient allocation of resources for population screening should focus on those tests which will divide the whole population into different levels of risk of handicap. This does not imply that only a very small high-risk group should then be placed under extra surveillance and the "at risk" register eliminated. (2 refs.) - *E. L. Rowan.*

National Children's Bureau 1 Fitzroy Square
London W. 1., England

3192 **PARISI, DOMENICO.** Development of syntactic comprehension in preschool children as a function of socioeconomic level. *Developmental Psychology*, 5(2): 186-189, 1971.

A test of syntactic comprehension was administered to 144 Ss aged 3 to 6.5 years in order to measure that ability as a function of age, sex,

and socioeconomic status (SES). Both age and SES were found to affect test performance. Sex did not influence performance either by itself or in interaction with the other 2 variables. The main interest in the results lay in the clear effect of SES on such a basic ability as comprehension of the syntactic structure of heard sentences. From the ages of 2 to 3 years, the importance of linguistic stimulation increases, and the culturally and linguistically impoverished condition of low-SES families increasingly hinders the language development of the child. A preschool education which does not develop the linguistic competence potentiality that is not realized at home, is inadequate. (10 refs.) - *B. J. Grylack.*

Instituto di Psicologia C.N.R.
Via C. De Lollis, 12
Rome, Italy

3193 New syllabuses of general education subjects for the auxiliary school. *Soviet Education*, 14(1,2,3): 90-96, 1971-1972.

In November, 1970, the Collegium of the U.S.S.R. Ministry of Education approved new syllabuses of general education subjects for auxiliary schools. These schools have the responsibility of correcting the defects in the development of cognitive processes in MR children and of resolving the problem of social adaptation faced by these children. The principle of accessibility of the content of instruction to the majority of pupils at each stage of instruction received strong consideration when the syllabuses were compiled. The experience of these schools over a period of many years has made it possible to define an optimal level of requirements for pupils in each general education subject. The overall amount of knowledge and skills required for success in each subject has remained virtually the same in the new syllabuses. The question of differentiation of requirements for various categories of children within each school subject merits further study. (1 ref.) - *B. J. Grylack.*

3194 **IAKOVENKO, M. I.** Job placement for auxiliary school graduates. *Soviet Education*, 14(1,2,3): 97-102, 1971-1972.

A composite study was done of 220 MR individuals who completed studies at an auxiliary school over an 18-year period. Of the total number, 166 had graduated from the school. Oligophrenia was diagnosed in 178 pupils, and the remaining 42 had other forms of mental disturbance. Only 47 of the Ss were employed in the job for which they had been trained in the school. Several Ss had been able to master another specialty after leaving school. Of the original 220 Ss, 194 are working full time, 7 are performing temporary seasonal work, and only 19 are not working. Job ratings have indicated that most of those employed are conscientious in their work. The joint impact of therapeutic and pedagogical measures during the period of instruction appears to exert a positive influence on the vocational training of these pupils. (No refs.) - *B. J. Grylack.*

Auxiliary School No. 93
Sverdlovsk, U.S.S.R.

3195 **TSUKERMAN, I. V.** Aids for children with multiple handicaps. *Soviet Education*, 14(1,2,3): 183-193, 1971-1972.

A general-purpose system enabling children with combinations of defects to transmit and receive alphabetic information with the aid of a special typewriter was used by 10 trainees. Five Ss had damage to the motor sphere accompanied by blindness, deafness, or speech defects, 1 child had no hands, and 3 were blind and had impairment of coordinated movement. The adult S was practically blind, and his hands had been amputated. The cybernetic communication devices which constitute the basic units are adjusted according to the specific handicap by replacement of 1 or more of the 12 individual units. The assignment given to the trainees was to memorize the cybernetic code and to achieve automatic skills in its transmission. In selection of the keyboard for each S, consideration was given to the movement of arms, prosthetics, and the range of their effect. A Braille keyboard was used for the blind Ss. Upon completion of the course, which ranged from 11 to 14.5 hours, all Ss converted rapidly to cybernetic code. (3 refs.) - *B. J. Grylack.*

U.S.S.R. Academy of Pedagogical Sciences
Moscow, U.S.S.R.

3196 **POTTER, HOWARD W.** Mental retardation: The cinderella of psychiatry. In:

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Menolascino, Frank, J. ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications 1971, 14-24.

Modern psychiatry has lost its original initiative in MR to other disciplines and must revitalize its interest and efforts in behalf of MRs. Historically psychiatry developed the MR treatment bases upon which present MR treatment rests. At the turn of the century psychiatry lost its leadership in MR due to the development of the mental test approach, the view of MR's inherent hereditary causes and the notion of irreversible brain damage as the source of MR. The modern psychiatrist is principally a behavioral scientist and must concern himself again with the assessment and treatment of the personality problems and learning impairments of MRs. Intensive psychotherapy can lead to striking improvements in MRs, especially EMRs. Efforts should be made both to reorient medical school training toward more education regarding MR and to provide psychiatric care for MRs and their families. (50 refs.) - *S. Glinsky.*

State University of New York Letchworth Village
Graduate Course in Mental Retardation
Thiells, New York

TREATMENT AND TRAINING ASPECTS – Psycho-social

3197 **PAUL, HOWARD A.; & MILLER, JOEL R.** Reduction of extreme deviant behaviors in a severely retarded girl. *Training School Bulletin,* 67(4): 193-197, 1971.

A combination of positive reinforcement and the time-out (T.O.) procedure developed by Zeilburger was successfully used to reduce the incidence of negative behavior in a 12-year-old institutionalized SMR girl. The child (IQ 23) displayed numerous self- and object-destructive behaviors, as well as bizarre psychotic-like behavior, including inappropriate laughter, smearing of feces, and disrobing. Previous efforts at nonbehavioral therapy were unsuccessful in reducing the child's negativism and failed to increase language production or interpersonal involvement. Hyperactivism and distractibility also persisted, despite chemotherapy. After 20 15-minute sessions of behavior modification therapy, held 3 times weekly, substantial amelioration of the negative behavior was noted. Positive behaviors – following directions, self-directed positive behavior, and communication – were rewarded with cereal and negative ones discouraged by the T.O. procedure. Once the positive behaviors are firmly established another resident in the school can be trained to maintain reinforcement within the daily activity program. (6 refs.) - *N. Mize.*

American Institute for Mental Studies
Vineland, New Jersey

3198 **HALL, R. V.; FOX, R.; WILLARD, D.; GOLDSMITH, L.; EMERSON, M.; OWEN, M.; DAVIS, F.; & PORCIA, E.** The teacher as observer and experimenter in the modification of disputing and talking-out behaviors. *Journal of Applied Behavior Analysis,* 4(2): 141-149, 1971.

"Talking-out" and argumentative behaviors displayed by students in regular and special education classes from the first grade to junior high school in White middle class and Black poverty areas were examined in order to seek the means to modify these behaviors. The study was carried out by teachers, who acted as both the experimenters and primary observers of the behaviors that were modified. The teachers used a combination of extinction for inappropriate talking and some sort of reinforcing event for appropriate behavior to bring about the desired change. The fact that talking-out is a problem of major concern to teachers is indicated by the fact that some form of this behavior was selected for modification by 23% of those enrolled in the behavior management classes which produced this study. It appears that teachers can develop effective observation and reinforcement procedures, can carry out experimental manipulations, and, therefore, can employ behavioral analysis as a tool in their classrooms. (13 refs.) - *B. J. Grylack.*

Juniper Gardens Children's Project
Kansas City, Kansas 66101

3199 **GARCIA, EUGENE; BAER, DONALD M.; & FIRESTONE, IRA.** The development of generalized imitation within topographically determined boundaries. *Journal of Applied Behavior Analysis*, 4(2): 101-112, 1971.

Two female and 2 male residents of an institution for the MR were used in a study to determine the limits of generalized imitation which may possibly be designated by reinforcement of a selected range of imitative response topographies. The nonimitative Ss were trained successively to imitate a number of different responses demonstrated by the experimenter, including small motor, large motor, and short vocal types of responses. The development of an imitative repertoire was observed in each S, although it was restricted topographically. The restriction was controlled by the recent training history of imitative response topographies in all 4 Ss. The multiple baseline technique demonstrated that imitation of similar topographical responses remained unaffected until training was instituted for responses representative of those types. (12 refs.) - *B. J. Grylack.*

University of Utah
Salt Lake City, Utah 84112

3200 **AZRIN, N. H.; & FOXX, R. M.** A rapid method of toilet training the institutionalized retarded. *Journal of Applied Behavior Analysis*, 4(2): 89-99, 1971.

Nine male incontinent and PMR hospital residents received training in which positive reinforcement was given for appropriate elimination in an attempt to accomplish rapid toilet training with these Ss and to help them achieve continence during the daytime in the ward setting. A "pants-alarm" and "toilet-chair" apparatus aided in the accomplishment of immediate detection, prevention of behavior incompatibility with proper habits, increase in the frequency of urination, stimulus control of elimination response, positive reinforcement of correct habits, and the independent toilet approach. During the first 7 weeks of the posttraining period, there was a 90% reduction in incontinence. Training required a median of 4 days and a mean of 6 days. Eventually almost all daytime incontinence was eliminated by the procedure. The importance of the program lay in its view of proper toileting as a complex and lengthy chain of responses that includes social, physical, and physiological stimuli and that requires strong positive and negative operant consequences rather than as a simple associative muscular reflex response to internal stimuli. (15 refs.) - *B. J. Grylack.*

Anna State Hospital
Anna, Illinois 62906

3201 **MARTIN, JERRY A.** The control of imitative and nonimitative behaviors in severely retarded children through "generalized-instruction following." *Journal of Experimental Child Psychology*, 11(3): 390-400, 1971.

Four institutionalized SMR males served as Ss in an experiment to determine whether unreinforced imitations could be maintained when interspersed among reinforced, nonimitative behaviors. For 2 Ss, nonimitative behaviors were reinforced while imitative behaviors were not. For 2 others, imitative behaviors were reinforced and nonimitative behaviors were unreinforced. Maintenance of unreinforced "instruction-following" was found to be dependent upon the response-contingent reinforcement of other "instruction-following" behaviors. Experimentation showed that unreinforced imitative behavior may be maintained in a manner other than by reinforcing other imitative behaviors. The data suggest the possibility that after imitative behavior is acquired it may not be controlled by the same stimuli which regulated it during acquisition. (21 refs.) - *B. J. Grylack.*

University of North Carolina
Chapel Hill, North Carolina

3202 North Carolina Council on Mental Retardation. *Foster Grandparent Project*, Western Carolina Center, Morgantown, North Carolina, 1968. 16 p.

The Foster Grandparent Project at Western Carolina Center for the retarded is described. It began as a 1 year project funded by a grant from the Office of Economic Opportunity, and was to demonstrate the use of older persons of limited income in serving to meet emotional needs of the retarded children. Initially 38 grandparents were

paired with 50 children, to whom they gave patience, closeness, and understanding. Weekly meetings were held between staff and grandparents to iron out difficulties. Quarterly data-gathering interviews were carried out by a social work supervisor. Many of the children's parents had positive responses to the program and therefore became more involved in it. It was one of the most constructive and rewarding programs the Center ever had, since both the children and older people benefited. (No refs.) - *T. C. Turner.*

North Carolina Council on Mental Retardation
Raleigh, North Carolina

3203 **DONOGHUE, E. C.; & ABBAS, K. A.** Unstable behaviour in severely subnormal children. *Developmental Medicine and Child Neurology*, 13(4): 512-519, 1971.

Senior ward staff completed questionnaires describing behavior problems of 293 SMR children in residential care. Of the 293 children, 148 were judged to have some type of disturbed behavior; 103 showed 2 or more disturbed behaviors. Commonest behavior problems were excessive attention-seeking, overactivity, and excessively noisy behavior; other problems were aggression, destruction, wandering, escaping, and self-mutilation. More males than females showed disturbed behavior. In particular, excessive attention-seeking and overactivity were more frequent among males; the other types of disturbed behavior did not show any sex bias. The least disturbed patients were those with cerebral palsy. An inverse relationship was found between frequency of major epilepsy and disturbed behavior. Significantly more of the Ss who were visited less than once in 6 months were disturbed than of those who were visited once a month or more ($x^2=8.48$, $p<0.01$). No significant relationships were found, however, between frequency of disturbed behavior and medical diagnosis, IQ, domicile prior to admission, or number of years hospitalized. (14 refs.) - *K. L. Woodworth.*

Queen Mary's Hospital for Children
Carshalton, Surrey, England

3204 **BALTHAZAR, EARL E.; ENGLISH, GEORGE E.; & SINDBERG, RONALD M.** Behavior changes in mentally retarded children following the initiation of an experimental nursing program. *Nursing Research*, 20(1): 69-74, 1971.

MR children involved in an experimental nursing program demonstrated varying behavior changes. A combination of nurturant nursing and conditioning principles was used to develop individualized nursing care programs for a random sample of 8 SMRs showing self-destructive and disturbed behaviors. A randomly selected group of 8 controls remained under the usual care. Testing with the Central Wisconsin Colony scales of adaptive behavior before and approximately 3 to 4 months after treatment revealed significant SMR and control group differences. These included the 3 social behavior factors of unskilled verbalization, failure to respond to contact by others, and passive response to contact by others. Although a trend existed toward improved nonverbal communication, there were no significant differences in resistive responses to others, posturing and stereopathy, or inappropriate responses to others. (8 refs.) - *S. Glinsky.*

Central Wisconsin Colony and Training School
Madison, Wisconsin

3205 **COLLINS, DEAN T.** General principles of training programs for the mentally retarded. In: Menolascino, Frank J. ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 371-378.

Training programs for MRs must be based on general principles whose importance cannot be overemphasized. These principles include the view of the MR as a human being first and only incidentally as an MR, the changing nature of program planning, and the necessity of both a total life plan as well as of immediately attainable goals. The physician must foresee developmental crises and prepare the parents for them, utilize the simplest possible services and regard institutionalization as only one phase of the MR's complete life plan. It is unwise to outline a single training program for all MRs, due to the ever-changing nature of each MR's requirements. (4 refs.) - *S. Glinsky.*

The Menninger Clinic
Topeka, Kansas

TREATMENT AND TRAINING ASPECTS – Occupational

3206 Employment of the mentally handicapped (retarded). *Journal of Rehabilitation in Asia*, 12(2):36, 1971.

Employers involved in the Institute of Industrial Launderers' 3-year training program for MR workers have given their retarded employees high marks for safety, attendance, and the ability to accept repetitive work. Quantity and quality of production were rated high in 57%; 46% were regarded as rapid learners, while an additional 32% learned more slowly, but were readily able to apply their training skills to the actual work situation. Other industrial employers surveyed similarly rated the work records of their retarded employees as comparable to those of other non-retarded workers on the same jobs. In 10 years the number of successful MR rehabilitations has increased from 2,000 to 19,000 (by 1968). (No refs.) - *N. Mize.*

3207 **MALIN, FR. A.** Sheltered employment for the handicapped in America. *Journal of Rehabilitation in Asia*, 12(2): 32-35, 1971.

Sheltered workshops for the MR in the United States, while they have recently been greatly increased in number, are still at a relatively early stage of conceptual development. Criticized as generally unrealistic and low in efficiency when compared to similar vocational training programs in Europe, there are nevertheless several innovative features in the American programs. Despite social handicaps of trade union antagonism and the trend toward increased automation, advances in the use of "half-way" and "quarter-way" houses to maximize employment opportunities have proven particularly successful. In the half-way house adult retarded live in an urban residential setting, near to suitable employment, and are under care of a guardian who helps with job placement and acts as counselor to the residents. The quarter-way concept provides even closer supervision by segregating the employable adolescent MR in a pre-placement cottage life program of social and vocational counseling while they are being trained in job skills at a nearby sheltered workshop. Sheltered work farms are being pioneered in the southern U.S., and in other areas various occupational arrangements, sometimes involving extensive private industry and government cooperation, are being implemented. Sheltered workshops have contributed much to successful rehabilitation of the MR, and their many proven benefits to the individual and to society alike warrant careful evaluation and program expansion. (16 refs.) - *N. Mize.*

3208 **SEILER, JOSEPH.** Preparing the disadvantaged for tests. *Vocational Guidance Quarterly*, 19(3): 201-205, 1971.

Disadvantaged persons usually perform poorly on employment tests because the tests have built-in biases severely limiting the performance of culturally deprived and undereducated individuals. The results of some E&D projects exploring the possibilities of preparing hard-to-employ individuals to take jobs are reported. Methods are discussed which aid applicants to overcome their fear of taking tests. It has been found that pretesting orientation can improve the test-taking capabilities of the disadvantaged. (No refs.) - *T. C. Turner.*

U. S. Department of Labor
Washington, D. C.

3209 **SALOMONE, PAUL R.** A client-centered approach to job placement. *Vocational Guidance Quarterly*, 19(4): 266-270, 1971.

An argument is presented for the client-centered job placement method rather than the "selective placement" approach as the key to successful placement of vocationally handicapped persons. Three main points are made: that the traditional placement approach is inefficient and ineffective; that a client-centered approach is most growth-facilitating for the client; that the key to successful placement is the counselor's optimistic attitude that the client will get a suitable job within

a time period agreed upon. The client-centered approach is then discussed at some length and the keys to placement success summarized. (13 refs.) - *T. C. Turner.*

Syracuse University
Syracuse, New York

3210 **RISCH, FRANK; & SAMUELS, SOLON D.** Gainful employment: A psychiatric prescription. *Journal of Rehabilitation,* 37(2): 22-25, 1971.

The potential value of the rehabilitation workshop as a means for restoring social and economic status to the poorly adjusted neuropsychiatric person and aiding him in meeting the normal standards of cooperative living in the community is described. Some observations on the psychiatric aspects of work are made and 3 cases discussed in illustrating the observations. It is noted that the rehabilitation workshop constitutes a good testing ground to discover the kind of jobs that best suit the emotionally troubled patient, enabling him to become better socially adjusted and eventually to enter the mainstream of social living. (2 refs.) - *T. C. Turner.*

Veterans Administration Center
Los Angeles, California

3211 **ROSSIT, EDWARD A.; & GRAYBEAL, EARL R.** Sheltered workshop certification. *Journal of Rehabilitation,* 37(3):30-31, 1971.

In 1970 in the state of Washington there were 50 sheltered workshops. Since they received state and federal funds, it became necessary to examine the quality of these facilities by inspecting them in order to "accredit" or "certify" them as conforming to adopted standards. The manner in which the survey was conducted is reported and recommendations are made for future surveys of this type. (No refs.) - *T. C. Turner.*

No address

3212 **CHAFFIN, JERRY D.; SPELLMAN, CHARLES R.; *REGAN, C. EDWARD; & DAVISON, ROXANA.** Two followup studies of former educable mentally retarded students from the Kansas Work-Study Project. *Exceptional Children,* 37(10): 733-738, 1971.

In 1967 and 1969 2 followup studies were made of students who had participated in the Kansas Work-Study Project (1964-67) and a comparison group from other school districts in an attempt to evaluate the usefulness of the program in post-school adjustment. The 3-year project's main purpose was to provide comprehensive rehabilitation, diagnosis and evaluation of employment potential, actual job tryout and training, job placement supervision, and curriculum activities for educable mentally retarded (EMR) students with IQs from 45-80 and 3 or more years retarded achievement. Results of the 1967 followup study indicate that the experimental group had more Ss who graduated and fewer who dropped out of school. Sixteen of 17 (94%) of the experimental graduates as opposed to 4 of 7 (57%) of the comparison graduates were employed, in training, or housewives. Results of the 1969 study indicate that more experimental than comparison Ss graduate from high school, and vocational adjustment of the 2 groups was nearly the same. Results suggest that at least three-fourths of the EMR are capable of some kind of employment without the necessary preparation of a work-study program. A work-study program seems to enhance the employability which already exists for the students rather than simply make them employable. (15 refs.) - *T. C. Turner.*

*Shawnee Mission School District No. 512
Shawnee Mission, Kansas

3213 **BROWN, LOU; JOHNSON, SYLVESTER; GADBERRY, EVE; & FENRICK, NANCY.** Increasing individual and assembly line production rates of retarded students. *American Institute for Mental Studies Training School Bulletin,* 67(4): 206-213, 1971.

Six TMR (ages 15-20, IQs 38-67) showed increased work efficiency and production rates in envelope stuffing as a result of manipulation of work arrangements (individual and assembly line) and reinforcement (social only and social plus tangible). Of 2 procedures (individual production, in which one person completes all task components, and assembly-line production, in which

each S is responsible for only one task component), the first induced consistently higher production rates (rates increased also with use of tangible reinforcements keyed to attaining a specific goal). Work was performed at tables in a public-school classroom in 15-minute work periods. Instructional objectives were defined so that behavioral components could be directly measured. It is evident that retarded students respond to work-environment changes. (5 refs.) - *B. Berman.*

University of Wisconsin
Madison, Wisconsin

3214 **O'TOOLE, RICHARD; & CAMPBELL, JOHN.** A situational approach. *Journal of Rehabilitation*, 37(4): 11-13, 1971.

The work adjustment process is conceptualized in terms of role theory and as the process of socialization whereby individuals attain knowledge, skills, and dispositions enabling them to work well as effective members of groups. Adult socialization takes place in great part by the individual learning what is required for participation. Given a desire to participate, the ability to assess and meet demands of a situation, the individual turns himself into the kind of person demanded by the situation. This is known as a situational adjustment. In order to have a work setting for training the client to learn to adjust to certain situations, a rehabilitation counselor must have a work setting and support. A rehabilitation workshop is authentically patterned after industry, providing adjustment situations similar to those in industry. Role relationships are learned and practiced; work attitudes, organization of work, and performance levels are learned. The work adjustment counselor analyzes situations and uses them to direct the situational adjustment process. He must be supported by the organization or agency. Organizational programs must be flexible to adapt to the client's needs. Also important is the agency's ability to provide support while the client is in training, which reduces role conflict. The positions the agency can develop for its clients' placement after training are important, as they can provide reward and motivational value to clients. (5 refs.) - *T. C. Turner.*

Vocational Guidance and Rehabilitation Service
Cleveland, Ohio

3215 **KRANTZ, GORDON.** Critical vocational behavior. *Journal of Rehabilitation*, 37(4): 4-16, 1971.

Work adjustment as a means for promoting critical vocational behavior is focused upon. The positive behaviors a person must have in order to become and remain employed are: job objective behaviors, job-getting behaviors, and job keeping behaviors. There are also critical employment-coupled behaviors: social, community, and general, and personal living competencies. These behaviors can be analyzed from any one of a number of different theories. However, a checklist is only a tool and human nature is flexible enough to defy tidy lists. But rehabilitation has developed the ability to deal with unique individuals and situations. (1 ref.) - *T. C. Turner.*

Co-operative School-Rehabilitation Center
Minnetonka, Minnesota

3216 **CAMPBELL, NORMA.** Techniques of behavior modification. *Journal of Rehabilitation*, 37(4):28-31, 1971.

Techniques of behavior modification based on operant conditioning were used to develop the work potential of MR and other handicapped. Treatment was begun by: objectively describing the present and wished-for (target) behavior; recording rate of frequency of behavior being studied; determining behavior consequences; modifying events following undesirable behavior. Various techniques are: use of automated equipment (so that clients could monitor their own performance); token reinforcement (productivity increased because clients could exchange tokens they earned for bus trips, tours, and so forth); work-contingent reinforcement; and isolation-avoidance procedures. Behavior modification/operant conditioning techniques are not the only method of helping handicapped clients, but they allow objective measurement of behavior and lend themselves to evaluation as a treatment process. (10 refs.) - *T. C. Turner.*

No address

3217 **AYERS, GEORGE E.** Counseling in work adjustment programs. *Journal of Rehabilitation*, 37(4): 31-33, 1971.

The utility of counseling in work adjustment programs is discussed along with some techniques. Individual counseling assesses behavior and suggests alternate patterns, guiding the client in peer and supervisor interaction. The counselor can orient the client to his new work setting, interpret adjustment and employment goals, interpret standards of work behavior and production goals, and participate in planning further beneficial services. Questions have been raised regarding the value of this kind of counseling, but it can be used to help a person through trauma and is necessary with certain clients. "Video-counseling" (videotaping samples of client behavior) can provide feedback to the client and is very useful in modifying behavior and attitudes. It can be used in improvement of performance and development of social skills. Modeling—the systematic observation of desired behaviors—can be extremely effective in transmitting entire behavior repertoires. Group counseling provides a new and supportive reference group for its members; old behaviors are examined and feelings of self-worth may be enhanced by group acceptance. It is particularly effective where individual clients have problems of an interpersonal nature. It is also useful in working with disadvantaged clients. (22 refs.) - *T. C. Turner.*

Mankato State College
Mankato, Minnesota

3218 **HOGAN, GAIL; & *BUTTON, WILLIAM H.** An empirical perspective. *Journal of Rehabilitation*, 37(4): 16-19, 1971.

The authors' research efforts have been directed at developing operational procedures able to measure flows of client behavior in sheltered workshops in order to assess capacity for performing production work. Two bodies of data exemplifying this approach are cited. The first was from studies of 2 workshop clients whose prognoses were very different. One, who was unable to remain continually in a work role, also had poor measurement of productive behavior. The other, whose rehabilitation was successful, had stability and organized control across a variety of tasks. The second body of data concerns the behavior of 26 clients observed in workshops, half of whom were considered placeable, half of whom were not. Thirteen 15 minute observations were made. Separate measurement was made of the time duration during which the subject performed the job and during periods of delay. During working times, the number of job cycles repeated was recorded, providing a continuous measure of productivity. The placeable clients took the initiative to resupply their work stations more often than did the unrehabilitable; they also had fewer personal production delays and spent much more time in social interaction. The data indicate that ability to participate and control one's social contacts is a critical dimension of the rehabilitation process. The workshop is a realistic context in which to observe and measure work behavior so that client potential can be evaluated and appropriate techniques and strategies can be formed to help the client on his way to useful and independent living. (6 refs.) - *T. C. Turner.*

*Cornell University
Ithaca, New York

3219 **HOFFMAN, PAUL R.** Work evaluation and adjustment: The relationship. *Journal of Rehabilitation*, 37(4): 19-22, 1971.

There are things in common between work evaluation and work adjustment, but substantial differences between the two indicate that they are 2 different entities. They deal with the same behavioral variables—with difficulties encountered in the work situation—and are closely related to the counseling process. Work evaluation is concerned with a wider range of behavior factors than work adjustment; the two processes are administered differently and are perceived differently by counselors and clients. Work evaluation focuses primarily on assessment, while work adjustment focuses on behavior changing. They are not mutually exclusive. There is need for work evaluation before a client is referred to a treatment program in order that a realistic and successful program may be identified. The unemployability of the individual must first be determined; then he is referred to an adjustment program. If he becomes employable, evaluation of specific employability factors is determined. He is then referred either for training or into processing for placement. Work evaluation and work adjustment are two aspects of an overall process of matching the individual with the job. (5 refs.) - *T. C. Turner.*

Stout State University
Menomonie, Wisconsin

3220 **DUNN, DENNIS J.** Work and behavior change. *Journal of Rehabilitation*, 37(4): 22-25, 1971.

The effectiveness of work in producing behavior change is discussed. Work approaches to behavior change are experiential and rely upon client involvement and experiences in a specific setting; they rely upon 3 modes of behavior change: identification, kinesthetic experience, and mystical experience. Identification is a process by which the person, in contact with behavior models, learns to imitate behavior patterns. Kinesthetic experience is knowing by doing—the acquiring of job proficiency through experience and the self-esteem that comes with job competency. Mystical experience is a term used to classify behavior change which is not attributable to specific activities. This experience is rare in work adjustment programs and is usually related to religion when it occurs. The above are some of the underlying processes of change in the work adjustment program. Positive behavior change can occur, it is asserted, without direct intervention of professional staff, but a risk in using work alone to produce change is noted: inappropriate behavior can also be modeled and tasks can destroy self-esteem as well as build it. (13 refs.) - *T. C. Turner.*

Stout State University
Menomonie, Wisconsin

3221 **MINK, OSCAR G.** Learner-oriented instruction. *Journal of Rehabilitation*, 37(4): 25-27, 1971.

A learner-oriented teaching system is discussed as the most appropriate way to learn for clients who enter a work adjustment program with deficiencies and years of failure behind them. There are 3 key concepts in the program: (1) Laws of control, or a person's expectations regarding his own ability to achieve success from his environment. This has an influence on the nature of his goal oriented behavior. It is necessary to aid clients to develop higher internal control expectancy, since this will help in job proficiency and success. (2) Teaching and counseling, which enable the client to become more self-directed. (3) The mastery concept, or all the criteria for total success in a work adjustment program. The basic strategy advocated is systematic design of several vocational skill development steps which would end at a certain set of performance criteria representing a set of job skills for a given work role. The final result would be a highly effective learning environment. (14 refs.) - *T. C. Turner.*

Regional Education Laboratory
Durham, North Carolina

3222 **HOFFMAN, PAUL; & DUNN, DENNIS.** The practice of work adjustment. *Journal of Rehabilitation*, 37(4): 2, 1971.

A need was felt for bringing together in one place a resource for the practitioner of work adjustment, one which would provide a view of theory and techniques of work adjustment in the workshop setting, showing the current state of practice and educating in the specific techniques of work adjustment. The result was a special issue on work adjustment in *The Journal of Rehabilitation*. The articles cover various topics: the confusion-causing available literature on work adjustment, limited validation of techniques, little agreement on terminology, the common objectives characterizing adjustment programs, behavioral ingredients of employability and job success, work evaluation, the processes of change involved in work itself, uses of a learner-oriented instructional system, behavior modification techniques, and varied approaches to counseling. This issue constitutes not a comprehensive treatment but rather an overview intended to reflect the current practice of work adjustment. (No refs.) - *T. C. Turner.*

No address

3223 **ROSS, DONALD R.; & BRANDON, THOMAS L.** In pursuit of work adjustment. *Journal of Rehabilitation*, 37(4): 6-8, 1971.

A 4-step procedure was used to ascertain whether a work adjustment methodology could be developed from current materials: (1) a novice in the rehabilitation field attempted to review the literature and develop a working model; (2) experienced professionals analyzed the same literature and attempted to develop a working model; (3) these endeavors were summarized; (4) recommendations were made for future research in the

field. The novice encountered great gaps and lack of clarity in the literature, with lack of uniformity of definition or terminology. The professionals discovered two main theoretical positions on work adjustment, one describing conditions in which man and his job became congruent, one focusing primarily on the removal of unacceptable traits in the work personality which could limit successful placement. Two articles seemed to be the most useful for a rehabilitation supervisor: William Gellman's "The Vocational Adjustment Shop" and Malcolm R. Lowell's "Vocational Evaluation and Work Adjustment Services in Manpower Programs." The professionals also became frustrated in trying to arrive at a uniform methodology to be used in programs. As a result, recommendations are made for a project to be undertaken to develop an integrated work program based upon solid theory and research evidence. After development, this program should be carefully field-tested to prove its validity and usefulness. (11 refs.) - *T. C. Turner.*

University of Arizona
Tucson, Arizona

3224 **SANKOVSKY, RAY.** Adjustment services in rehabilitation. *Journal of Rehabilitation,* 37(4): 8-10, 1971.

Three basic types of adjustment services (personal, social, and work) in rehabilitation are discussed. Personal adjustment deals with personal problems hindering the individual's ability to deal effectively with others. A well structured personal adjustment program includes thorough evaluation of the individual's personal problems and an opportunity to change inappropriate behavior. Problem cases in social adjustment are usually recognized, but those individuals with unnatural dependency or poor communication skills tend to be overlooked. Their problems often stem from lack of experience or information. A program including classes in social skills, sex education, communication, and related areas is necessary. Work adjustment programs, as opposed to social adjustment programs, have received much attention. Work adjustment is a therapeutic process designed to enhance an individual's vocational potential skills in providing development of physical tolerance, and/or providing information or new experiences, and/or changing existing behavior. The workshop gives the opportunity to observe an individual in work situations and to guide him accordingly. (2 refs.) - *T. C. Turner.*

Auburn University
Auburn, Alabama

3225 **KAZDIN, ALAN E.** Toward a client administered token reinforcement program. *Education and Training of the Mentally Retarded,* 6(2): 52-55, 1971.

At Shore Training Center in Evanston, Illinois, 58 clients with mean CA of 29 (range 16 to 52) and median IQ of 40 (range untestable to 85) are employed in a sheltered workshop where they are grouped in 3 levels according to their ability to produce at their jobs relative to minimal industrial rates. In an attempt to promote work skills and habits in the interests of future competitive employment, a behavior modification program involving peers as reinforcers was implemented in the fall of 1968 in which 20 clients were reinforced with tokens for work performance and habits. Specific programs were initiated, using peer participation, to prompt normal conversation during the work day in 2 adult males (IQs 60 and 65) who were reluctant to talk with staff or peers, to encourage good posture in a brain-injured male (IQ 78), and to increase the interaction of an adult male (IQ 83), who often talked to himself, with his peers. Although the advantages achieved by employing clients as part of their own rehabilitation and that of their peers remains to be evaluated by research, it appears that involving peers in the treatment program whenever possible is desirable in reinforcement programs. (6 refs.) - *B. J. Grylack.*

Shore Training Center
Evanston, Illinois

3226 **HUSTON, DORA.** In Texas hiring the handicapped. *Rehabilitation Record,* 12(5): 33-35, 1971.

A program for selective placement of qualified handicapped people in State government positions has been started in Texas. Under the direction of the State Employment of the Handicapped Section of the newly organized Texas Rehabilitation Commission (TRC), the program

was responsible for placing at least 71 clients in 19 different agencies during its first 5 months. Since Texas does not have a civil service system for all employees, and the heads of many agencies are elected, implementation of the program depended upon securing the voluntary cooperation of the agencies involved. Each agency head was asked (1) to select a staff member to act as coordinator for the program; and (2) to write a letter to his field offices, if any, requesting cooperation. Responsibilities of the coordinator are to ensure that handicapped applicants have an equal opportunity for available employment for which they qualify, and to provide liaison with the TRC. Follow-up has included workshops for coordinators, a monthly newsletter noting participation and progress, a brochure, and an awards program to recognize achievement. Progress is assessed through a monthly reporting system. (No refs.) - *S. M. Rucker.*

No address

TREATMENT AND TRAINING ASPECTS – Therapy

3227 **BLACKLIDGE, VIRGINIA Y.; & EKBLAD, ROBERT L.** The effectiveness of methylphenidate hydrochloride (Ritalin) on learning and behavior in public school educable mentally retarded children. *Pediatrics,* 47(5): 923-926, 1971.

A double-blind, cross-over experiment to study the effects of methylphenidate hydrochloride (Ritalin) on 3 classes of EMR public school children has shown the drug to be effective in improving classroom behavior. A group of 31 Ss (19 male and 12 female), ranging in age from 9 to 14 years and in IQ from 51 to 88 (median = 70) participated in the study. Ritalin (10 mg) or a placebo was given at 9:00 a.m. and 1:00 p.m., Monday through Friday, after baseline evaluations were conducted. Evaluation after 4 mos (2 on Ritalin plus 2 on placebo), utilizing Burks Behavior Rating Scale, the Arithmetic section of Wide Range Achievement Test, Gray's Oral Reading Paragraphs (Form A), and Porteus Mazes (Vineland Revision), showed no significant improvement in learning could be demonstrated by the latter 3 tests; however, all 3 teachers reported improvement as measured by the Burks Behavior Rating Scale. Statistical analysis of results supported this conclusion. (11 refs.) - *M. S. Fish.*

Alameda County Mental Retardation Service
San Leandro, California 94577

3228 **HATTEN, JOHN T.; & HATTEN, PEQUETTI A.** A foster home approach to speech therapy. *Journal of Speech and Hearing Disorders,* 36(2): 257-263, 1971.

A 6-year-old child with limited language development was placed in a foster home for 5 days a week in a speech habilitation program. Therapy included an operant approach at the home and a 3-hour session every day at a university speech clinic. During his first week, 55 verbalizations were made and he spoke 5 separate words. During the last week, over 1,000 verbalizations were made and 65 words attempted. (8 refs.) - *T. C. Turner.*

University of Minnesota
Duluth, Minnesota

3229 The use of stimulant drugs in treating hyperactive children. *Children,* 18(3): 111, 1971.

Several conclusions of the report on hyperkinetic behavior disturbance in elementary school children are: (a) attention, learning, and social abilities are improved by certain amphetamines; (b) adequate diagnosis of a child may require use of psychological, educational, and social, as well as medical resources before treatment begins; (c) no evidence has been produced in 30 years of medical experience to show that treated children become drug abusers in later life. Further research into causes, prevention, and treatment of behavior disorders in children is necessary. The report also warns against permitting public attitudes toward the use of drugs to deprive

children of necessary medication. (No refs.) - *T. C. Turner.*

Office of Child Development
Washington, D. C. 20013

3230 **BOLL, THOMAS J.** Systematic observation of behavior change with older children in group therapy. *Psychological Reports*, 28(1): 26, 1971.

Systematic observation of group play therapy with multiply handicapped Ss (CA 3-11 years) is reported. Very young children became inhibited in play, vocalization, and speech when handled in a nondirective manner, but became less inhibited when in an atmosphere of emotional support and physical restraint. Older children treated by an active therapist did not noticeably increase their activity levels, but those seen by the nondirective, nonevaluative therapist increased markedly in their amount of play, vocalization, speech, and movement. (1 ref.) - *T. C. Turner.*

University of Washington
Seattle, Washington

3231 **SCHOPLER, ERIC; BREHM, SHARON S.; KINSBOURNE, MARCEL; & REICHLER, ROBERT J.** Effect of treatment structure on development of autistic children. *Archives of General Psychiatry*, 24(5): 415-421, 1971.

Autistic children tended to learn more adaptive behavior patterns in a structured than in an unstructured environment. Ss were 4 boys and 1 girl (CA range 4 to 8 yrs; SQ range 29 to 86) who had a clinical diagnosis of autistic or psychotic and functioned at the preschool level. The children were exposed to a relatively structured treatment program for 2 weeks (S_1) and then to a relatively unstructured program for 2 weeks (US_1); the cycle was then repeated (S_2, US_2). Changes in behavior from S_1 to US_1 were significant for attending ($p<.05$), appropriate affect ($p<.10$), relating ($p<.10$), and nonpsychotic behavior ($p<.05$). These behaviors were more appropriate in the structured situation. There was a tendency for appropriate behavior to occur more frequently during S_2. The amount of nonmeaningful vocalization was significantly lower during S_1 than during US_1 or US_2 ($p<.02$) or during S_2 ($p<.01$). Positive differences in behavior during the first cycle were generally not sustained when the cycle was repeated, and repetition of the cycle generally produced regression to the mean score. During the second cycle, a significantly greater number of adaptive responses occurred for relatedness during S_2 ($p<.05$). Children on a higher development level made more adaptive responses than children functioning on a lower developmental level. (13 refs.) - *J. K. Wyatt.*

University of North Carolina
Chapel Hill, North Carolina

3232 **BLOCKEY, N. J.** Aids for crippled children. *Developmental Medicine and Child Neurology*, 13(9): 216-227, 1971.

Aids for children aged 2 to 8 who have congenital spinal defects are not commercially available but can be made in the hospital workshop. Wheelchairs can be specifically developed for the severely disabled and walking can be encouraged by devices which provide stability and a firm grip, and prevent falling. Standing tables, hand-operated mobile aids such as converted chain-drive tricycles and direct drive mobile platforms, and walking aids give the disabled child confidence in his ability to move by himself. Most can be made relatively cheaply with steel tubing, casters, wheels, and parts of toys. (No refs.) - *E. L. Rowan.*

Royal Hospital for Sick Children
Oakbank, Glasgow C4, Scotland

3233 **LUCAS, ALEXANDER R.; & WEISS, MORRIS.** Methylphenidate hallucinosis. *Journal of the American Medical Association*, 217(8): 1079-1081, 1971.

Gross behavioral changes with hallucinosis were observed in 2 children and 1 adolescent who were receiving methylphenidate for symptoms characteristic of minimal brain dysfunction. A girl aged 6.5 years who had average intelligence received 90 mg of the drug over a 9-day period for hyperkinesia and distractibility, and she subsequently experienced catatonic withdrawal. A 10-year-old boy with severe learning and behavioral difficulties received 40 mg of methylphenidate over a 5-day period and suffered hallucinosis subsequently. A 15-year-old high-school girl

with a long history of hyperkinetic impulse disorder was seen following an episode of visual hallucinations after taking 80 mg of methylphenidate within 8 hours, an excess over her usual dosage. The most common side effect of methylphenidate intake is anorexia, which can generally be managed by dose adjustment. Stimulation and excitation may occur, although they are not the usual effects in children. Psychotic reactions, as seen in these 3 cases, are unusual. Used judiciously, methylphenidate and amphetamines are among the safest drugs for preadolescent children; however, they are not innocuous. (9 refs.) - *B. J. Grylack.*

951 E. Lafayette
Detroit, Michigan 48207

3234 **CLAGHORN, J.; NEBLETT, C.; SUTTER, E.; FARRELL, G.; & KRAFT, I.** The effect of drugs on hyperactivity in children with some observations of changes in mineral metabolism. *Journal of Nervous and Mental Disease*, 153(2): 118-125, 1971.

The finding that acetazolamide reduced cerebrospinal fluid (CSF) output by 50 to 60% with small but reproducible decreases in CSF potassium in 3 children with indwelling intraventricular cannulae prompted a double blind investigation of the calming effects of acetazolamide and amphetamine on 17 boys and 6 girls with a mean CA of 8 and mean IQ of 35, all of whom were diagnosed as familial or idiopathic retardates. Placebo and no treatment conditions were used for comparison. The behavior of the Ss was evaluated on the basis of performance in a "hyperactivity room" designed to obtain quantitative measures of their movements and 3 behavior observation rating forms. The active treatment condition in all cases was acetazolamide, effects on sleep being marked only for this treatment. Acetazolamide was found to reduce serum and red cell potassium, while no other electrolyte effects were observed with any drug. Of the 4 treatment conditions, acetazolamide (1-week administration) and placebo improved hyperactivity, with acetazolamide being statistically superior. In view of the many pharmacological actions associated with this drug, the theoretical significance of the definite reduction in hyperactivity noted in this short trial is unclear. (10 refs.) - *B. J. Grylack.*

Texas Research Institute of Mental Sciences
Houston, Texas

3235 **STEINBERG, GRACE G.; TROSHINSKY, CHARLES; & STEINBERG, HARRY R.** Dextroamphetamine-responsive behavior disorder in school children. *American Journal of Psychiatry*, 128(2): 174-179, 1971.

Ss were 46 children (CA 5-11 years) with behavior disorder and/or difficulty concentrating who were randomly administered either d-amphetamine or placebo for 4 weeks, given a 10-day vacation, and crossed over to the opposite condition for 4 weeks. Twenty-five Ss on d-amphetamine improved markedly. Those who improved most were Ss with at least one hard neurological sign or 2 or more soft signs. Although the drug produced a clear-cut behavior change, the authors do not feel that it is sufficient to institute a case finding and long-term treatment program. Rather, further prognostic studies are necessary. (6 refs.) - *T. C. Turner.*

Georgetown University Medical School
Washington, D. C. 20007

3236 **OBALDIA, MARIO DE; & BEST, GARY A.** Music therapy in the treatment of brain-damaged children. *Academic Therapy*, 6(3): 263-269, 1971.

The results of a program of music intervention in the education and treatment of 10 brain-damaged Ss (CA10-14 years) in a small, private clinic-school are reported. The 2-part program consisted of first having the Ss sing together in order to measure their rhythmic and melodic ability; secondly, they chose specific musical instruments to work with, then broke into 3 groups to learn piano, bongo drums, and guitar. The fourth group pursued interpretation of music, since all 3 Ss had played piano for some years. Each student practiced with the music therapist 5-10 minutes per day over 30 therapy sessions. Improvement in each S was noticeable regarding behavior change, attitudes toward society and academic subjects, and self-assurance. Music therapy seems to be a successful technique in treatment and training of brain-damaged children; it may be used with several different categories of

brain damage. It should, however, be given in conjunction with a good academic program when possible. (1 ref.) - *T. C. Turner.*

Penny Lane
Sepulveda, California

3237 **LARSON, THALIA.** Communication for the nonverbal child. *Academic Therapy,* 6(3): 305-312, 1971.

The Orange County (California) Developmental Centers for Handicapped Minors have been experimenting with a simultaneous sensory approach to motivate nonspeaking children to communicate verbally. It is actually a system of communication borrowed from the language of signs which is used with the deaf. At the same time the child sees the sign, he hears the word spoken and sees the object of the activity that the sign and word represent. The rationale for using sign language is that some children will be motivated to use body gestures in expressing ideas and may begin to verbalize when they learn the pleasure and power of communication. The technique may not be successful with all children, as some may be too retarded for training classes or may have a physical disability preventing oral language. Children who lacked environmental stimulation at the most propitious time for speech development and thus became silent may respond to a gesture language. They may then be able to go one step further and communicate verbally. The gesture language is intended to help stimulate oral language, not replace it. (No refs.) - *T. C. Turner.*

Special Education Schools
Orange County, California

3238 **WRIGHT, LOGAN.** Conditioning of consummatory responses in young children. *Journal of Clinical Psychology,* 27(3): 416-419, 1971.

Operant conditioning was successfully employed on 3 female MR Ss (2 to 6 years old) to induce them to accept either fluids or solid foods. Success was achieved within 3 weeks, by shaping or desensitizing Ss to consumption of various foods and fluids. A combination of social reinforcement and cognitive stimulation was especially successful in reducing resistance to consumption. (2 refs.) - *B. Berman.*

University of Oklahoma Medical Center
Oklahoma City, Oklahoma

3239 **TOLMAN, R. L.** "all life is a risk...". *Expectations,* 1(2): 4, 1971.

If physicians caring for neurologically injured children do not explore every possibility for cause and treatment, do not keep up with their reading or gamble on new treatment which might help the patient – in short, try to avoid risks by staying within the boundaries of what is already known and reliable – then these physicians are irresponsible. In many cases new treatments and approaches are helpful, and a certain degree of improvement may be noted in a particular child. In many situations the decision to avoid taking risks constitutes a risk in itself. (No refs.) - *T. C. Turner.*

3240 **ENGH, ROHN; & ENGH, JERI.** Editorial. *Expectations,* 1(2): 5, 1971.

Three issues central to the issue for health care for brain-handicapped children are discussed: (1) that there is a cure for the retarded brain; (2) that parents have an unrealistic goal of wanting their child to become "normal"; (3) that reliable counselors are needed in sifting out the swindle from the "new cure." Firstly, not many treatments title themselves "cures," although they may be helpful if used in conjunction with existing therapies. Secondly, reliable counselors are rare since most health professionals have an overburdened workday and are unable to read and assimilate new literature. Thirdly, most parents are quite realistic and sensible about their retarded child, realizing that he will never be "normal," and they are unwilling to cut their children off from growing to their individual potentials. Some children cannot be helped, but a child must not be condemned before he has had the benefit of screenings and treatment. Open dissemination of information to parents has been blocked for too long. It is strongly felt that this information must not be kept from those who could benefit from it, just for the sake of a few parents who might "chase rainbow cures." (No refs.) - *T. C. Turner.*

No address

3241 **SCHWARTZ, MARTIN L.; PIZZO, SALVATORE V.; & McKEE, PATRICK A.** Minimal brain dysfunction and methylphenidate. *New England Journal of Medicine*, 285(5): 293, 1971. (Letter)

It is disturbing that CIBA, on the basis of only meager data, has embarked on skilful advertising of Ritalin (methylphenidate) in management of children with minimal brain dysfunction. Although Ritalin might be useful in very rare cases of hyperkinesis, this appears in only a very small percentage of children with minimal brain dysfunction. Exploration of this pathology requires coordinated services of a professional group rarely available outside a medical center. (3 refs.) - *B. Berman.*

Duke University Medical Center
Durham, North Carolina

3242 **SALWA, BARBARA I.** Preliminary considerations in psychotherapy with retarded children. *Journal of School Psychology*, 9(1): 12-15, 1971.

Preliminary considerations in psychotherapy with MRs emphasize the responsibility which the school psychiatrist may have to deal with such cases. The MR child enters the therapeutic situation with a language deficit, emotional attitudes concerning MR, and social experiences. Suggestions are made for the successful implementation of group and individual therapy. (12 refs.) - *S. Glinsky.*

204 Cedar Crest Drive
Cedar Falls, Iowa 50613

3243 **WORK, HENRY H.** Insight into treating parents of the retarded. *Medical Insight*, 3(9): 52-53, 56-59, 1971.

Psychiatrists must be aware of a varied range of problems in treating the parents of MR children. Parents must be helped to reconcile hopes and plans for the child with reality and to accept the diagnosis without seeking a more hopeful one elsewhere. Problems to be countered include the parental tendency to isolate themselves from society, and blame projection within the family. Guilt feelings, which hinder the constructive use of outside resources, and the externalization of anxiety are further common factors with such parents. Since parental expectancy applies both to plans for the parents themselves as well as for the child, both sets of expectancies must be considered. (No refs.) - *S. Glinsky.*

No address

3244 **GILLETTE, HARRIET E.** Presidential address to the American Academy for Cerebral Palsy. *Developmental Medicine and Child Neurology*, 13(4): 533-535, 1971.

The development of the cerebral palsied child may be hindered in infancy through lack of an adequate amount of external stimulation (visual, auditory, tactile, and motor), and in childhood by lack of normal interaction with peer groups and the environment and by lack of appropriate intellectual stimulation and challenge. In adulthood, while the normal person is enjoying a career and/or family life, the cerebral palsied is undergoing the attrition of later stages of development. The deprivations of early years combine with the natural process of aging, resulting in poverty of thought and stimulation. To prevent this, management during his important develop mental years should include careful observation, objective testing, and realistic planning of long-range goals. Too often there is lack of integration among the specialties interested in the cerebral palsied. Goals of comprehensive care involve health, education, and welfare, and there must be united effort in these 3 areas. Physicians must increase their awareness of the social science aspects of chronic illnesses and utilize the support of other agencies and organizations. There must be a cooperative effort of many disciplines in evaluation, goal-setting, and implementation to achieve optimal management of the cerebral palsied. (No refs.) - *K. L. Woodworth.*

University of Colorado Medical Center
Denver, Colorado 80220

3245 **BEGELMAN, D. A.; & HERSEN, M.** Critique of Obler and Terwilliger's "Systematic desensitization with neurologically impaired children with phobic disorders." *Journal of Consulting and Clinical Psychology*, 37(1): 10-13, 1971.

The conclusions of Obler and Terwilliger's report (1970) describing an experimental behavior therapy technique are challenged. Obler and Terwilliger stated that their technique was effective in inducing neurologically impaired children with phobic disorders to confront their fear-inducing stimuli. A major criticism is that several behavior modification techniques have been confused by the authors, and they failed to be explicit about the role of the therapists. In measuring treatment success they arbitrarily differentiated aware and unaware Ss on the basis of a median split at the WISC IQ level of 75. They do not document the assumption that an isomorphic relationship exists between IQ and awareness, nor do they anchor their conception of the latter to any stimulus - response dimension. Their reliance on parents' verbal ratings as a dependent variable introduces further methodological difficulties. They do not present ordinal or interval change scale scores between pretreatment and posttreatment measures, nor do they indicate what steps were taken to establish consistency of the experimental procedure. Investigators carrying out future empirical tests of behavioral techniques must present clearly articulated and replicable therapeutic operations and provide objective measures of the behavior anomaly in question. (25 refs.) - *T. C. Turner.*

Veterans' Administration Center
Jackson, Mississippi 39216

3246 **STERNLICHT, MANNY; SIEGEL, LOUIS; & DEUTSCH, MARTIN R.** Evaluation of a remotivation program with institutionalized mentally retarded youngsters. *American Institute for Mental Studies Training School Bulletin*, 68(2): 82-86, 1971.

A study was conducted to elicit objective data concerning the effectiveness of remotivation procedures on SMR and PMR children and the attendants carrying out the program. Ss were 3 motivator-attendants and 24 SMR and PMR children (IQ 6-30, CA1-9 years), divided into 3 groups meeting in half-hour sessions 3 times a week for 3 months. They were evaluated 4 times by the Kuhlman Test of Mental Development (KTMD), the Vineland Social Maturity Scale (VSMS), Adjective Check List (ACL), and Behavior Rating Scale (BRS). Significant differences in group behavior occurred only on the BRS, but notable improvements in Ss' personal characteristics or verbal behavior were not noticed. However, the relatively short duration of the program and the fact that remotivation is not an effort to improve personal and verbal behavior per se are possible reasons for this. The remotivation technique, when employed with SMR and PMR children, is most effective in motivating them to be more sociable, alert, and responsive to their surroundings. The attitudes of the attendants were extremely positive during the study. (3 refs.) - *T. C. Turner.*

Willowbrook State School
Staten Island, New York

3247 **FOX, MAURICE S.; & LITTLEFIELD, JOHN W.** Reservations concerning gene therapy. *Science*, 173(3993): 195, 1971. (Editorial)

A realistic appraisal is made of the potential and liabilities of gene therapy. Possibilities of gene therapy under discussion include: introduction of DNA or chromosomes, either directly or by cell fusion; transfer of genetic particles from one host to another by virus-like particles containing DNA of the host cell; infection with active or inactive virus containing genes that can determine some particular biochemical function; or infection with a viral nucleic acid to which some cellular gene has been coupled. Only a small fraction of newborns' genetic disorders could be amenable to intervention by these techniques. Genetically dominant disorders, multigenic traits, and disorders resulting from extra chromosomes are not amenable to treatment by any of the above techniques. It also does not seem possible that recessive inborn errors of metabolism could be helped. There are certain hazards: many of the procedures are mutagenic and many viruses under consideration as vectors may produce tumors. Proponents of gene therapy for the most part ignore its hazards. Understanding of human genetics is still in a descriptive phase, with little, if any, knowledge of how to intervene safely. The authors hope that we will not, therefore, through misuse of gene therapy, do to ourselves what we have done to our environment. (No refs.) - *T. C. Turner.*

Massachusetts Institute of Technology
Cambridge, Massachusetts

3248 **TERWILLIGER, ROBERT F.; & OBLER, MARTIN.** Comment on research in behavior therapy. *Journal of Consulting and Clinical Psychology,* 37(1):14-15, 1971.

Response is made to Begelman and Hersen's (1971) comments on the authors' (1970) paper. (a) It is realized that the use of IQ as an awareness measure is open to criticism. It was included primarily to forestall any criticism that therapeutic failure was due to low verbal and conceptual abilities of neurologically impaired patients. (b) The dependent variables of the study were called inadequate. However, the therapist had direct observation of the subjects' willingness to interact with the phobic object and observed the behavior as having changed. There was no parental "debriefing." (c) The behavior modification technique was criticized as being unclear and unorthodox. The authors wished to show that it was possible to treat these symptoms but that the nature of the patient population required some revision in the standard procedure of systematic desensitization. The aim was not *in vivo* desensitization per se, but to do therapy on brain-injured patients. (3 refs.) - *T. C. Turner.*

New School for Social Research
New York, New York 10011

3249 **KIRCHER, ALFRED S.; PEAR, JOSEPH J.; & MARTIN, GARRY L.** Shock as punishment in a picture-naming task with retarded children. *Journal of Applied Behavior Analysis,* 4(3): 227-233, 1971.

An experiment to examine the effectiveness of electric shock as punishment for inattentive behavior and incorrect responses in a picture-naming task with SMR children (also using a no-shock situation for comparison) suggested that electric shock reduced inattentive behavior, somewhat increased correct responses, and somewhat decreased the ratio of errors to correct responses. Increasing the ratio of undesirable behavior to shock decreased differences between shock and no-shock procedures. (19 refs.) - *N. Jarvis.*

University of Manitoba
Winnipeg, 19, Manitoba, Canada

3250 **VUKELICH, RON; & HAKE, D. F.** Reduction of dangerously aggressive behavior in a severely retarded resident through a combination of positive reinforcement procedures. *Journal of Applied Behavior Analysis,* 4(3): 215-225, 1971.

A program of behavior therapy based on positive reinforcement procedures produced enduring changes in aggressive behavior of an 18-year-old inst SMR girl (IQ 14). The patient was restrained almost constantly in a timeout chair; upon being released, she would grab or choke attendants and ward members. In one experiment, she was subjected to timeout for choking responses only; upon being released she was provided with extra attention. Choking responses were followed by a return to the timeout chair; grabbing movements were countered with responses incompatible with them. After a week of this treatment, attention followed only upon responses incompatible with aggression and responses that were designed to produce attention. Timeout was now provided for both grabbing and choking responses. Finally, all extra attention was discontinued. A second experiment attempted to increase the amount of attention given the subject when she was unrestrained. Results of the program indicated a reduction in number of both choking and grabbing responses which is related to the factor of extra attention: when extra attention was discontinued, S averaged 4 aggressive responses a (12-hr) day; when periodic extra attention was introduced, she averaged less than one aggressive response every 6 days. Results suggest that programs relying upon positive reinforcement rather than aversive techniques will be most successful with MR when extra positive reinforcement is employed. (14 refs.) - *N. Jarvis.*

Anna State Hospital
Anna, Illinois 62906

3251 **CORTE, HENRY E.; WOLF, MONTROSE M.; & LOCKE, BILL J.** A comparison of procedures for eliminating self-injurious behavior of retarded adolescents. *Journal of Applied Behavior Analysis,* 4(3): 201-213, 1971.

Behavior therapy for self-injurious behavior (face slapping, banging, and scratching; hair pulling and finger biting) in inst MR adolescents utilized 3 approaches: extinction through elimination of all social consequences, reinforcement of non self-injurious behavior, and electric shock punishment. Results indicated that punishment was

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most effective, reinforcement next most successful, and extinction least effective. Since the effects of punishment were specific to the setting, it appears that to be successful it must be carried out in the many settings in which the self-injurious behavior occurs. (16 refs.) - *N. Jarvis.*

Plymouth State Home and Training School
Northville, Michigan 48167

3252 **HORNER, R. DON.** Establishing use of crutches by a mentally retarded spina bifida child. *Journal of Applied Behavior Analysis*, 4(3): 183-189, 1971.

Operant conditioning was used to teach a 5-year-old MR child with spina bifida to walk with crutches. Two weeks after treatment, which consisted of baseline, teaching the child to pull himself up on parallel bars, extinction, re-establishing use of parallel bars, baseline on use of crutches, establishing use of crutches, and follow-up, the child was walking to and from most activities. Since the study involves a single subject, no claims are made as to the general usefulness of the techniques in this condition. (4 refs.) - *N. Jarvis.*

State Home and Training School
Wheatridge, Colorado

3253 **MAHONEY, KURT; VAN WAGENEN, KEITH; & MEYERSON, LEE.** Toilet training of normal and retarded children. *Journal of Applied Behavior Analysis*, 4(3): 173-181, 1971.

A method of toilet training that instills and reinforces a sequence of behaviors leading to elimination was successful with 7 of 8 incontinent children, 5 of whom were MR. Each child was trained to initiate the routines of going to the toilet in response to an auditory signal, reinforced by physical and auditory prompts, social praise, and food. As the behavior began to be established, the reinforcers were reduced until the child responded to the auditory signal alone. Finally the child was able to carry out the total sequence of toilet elimination without signals or prompts. Aspects of the procedures that did not require the signalling device were then taught to the parents for generalization at home. (7 refs.) - *N. Jarvis.*

Arizona State University
Tempe, Arizona

3254 **AZRIN, N. H.; BUGLE, C.; & O'BRIEN, F.** Behavioral engineering: Two apparatuses for toilet training retarded children. *Journal of Applied Behavior Analysis*, 4(3): 249-253, 1971.

Toilet training of 4 PMR girls (ages 3-6) utilizing a toilet-chair apparatus and a portable pants-alarm apparatus to signal wetting of the pants proved nearly 100% successful after 3 days of training. When training was interrupted, wetting behavior resumed but disappeared when training was reinstituted. Both devices are inexpensive (about $10) and easily available. (10 refs.) - *N. Jarvis.*

Anna State Hospital
Anna, Illinois

3255 **GODDARD, P. J.; & BECKET, A. J.** A simple safety valve for infant resuscitator and ventilator gas circuits. *Lancet*, 2(7724): 584, 1971.

A spring-loaded blow-off valve has been devised for use in giving newborns intermittent positive-pressure ventilation. The valve, constructed mainly of Duraluminium, works in any position, is easily sterilized, can be set to blow off at any predetermined pressure within a reasonable range, and can be incorporated into any resuscitation equipment. (No refs.) - *B. Berman.*

Hammersmith Hospital
London, England

3256 **SMITH, BARRY T.; & MASOTTI, R. E.** Intravenous diazepam in the treatment of prolonged seizure activity in neonates and infants. *Developmental Medicine and Child Neurology*, 13(5): 630-634, 1971.

Intravenous injection of diazepam was used in 40 episodes of prolonged seizure activity occurring

in 21 neonates and infants. A solution of 1 mg diazepam/ml was administered by slow intravenous injection over a 2-minute period or until seizure manifestations ceased or side effects occurred. A total of 35 episodes (87%) was treated effectively in this manner, with the effective dose ranging from 0.08 to 2.72 mg/kg and averaging 0.68 mg/kg. Respiratory arrest occurred in 1 child who had ventricular septal defect and pulmonary hypertension, which required assisted ventilation. Parenteral diazepam diluted to a 1 mg/ml solution and administered slowly intravenously is a safe and effective treatment for prolonged seizure activity in neonates and infants. The wide dosage range found to be effective here underlines the importance of utilizing the intravenous route with clinical titration. (11 refs.) - *B. J. Grylack.*

Queen's University
Kingston, Ontario, Canada

3257 **KUDRIAVTSEVA, V. P.; & FOMCHENKOVA, L. F.** Therapeutic speech training for oligophrenics. *Soviet Education*, 14(1,2,3): 166-173, 1971-1972.

Therapeutic speech training was conducted for 10 MR children whose defects were caused by organic brain injury dating from infancy or early childhood. The children were able to pronounce complex phrases with phonetic perfection and without grammatical errors, but their speech was imitative. They could also memorize verses, but with a poor understanding of the content. In order to assist the children in the formation of spatial concepts and in personality development, particularly in development of the differentiation between self and others, the patients' attention was concentrated on kinesthetic sensations. An understanding of various somatic movements was developed in the children by directing their attention to the actions while they were being performed. Following instruction, the children were able to distinguish the various movements in a more specific manner. It appears that special correctional exercises, as part of a complete therapeutic course, have a positive impact both on general conduct and speech. (No refs.) - *B. J. Grylack.*

U.S.S.R. Academy of Medical Sciences
Moscow, U.S.S.R.

3258 **UL'IANOVA, T. K.** Certain techniques of speech training for auxiliary school pupils who are learning about their environment. *Soviet Education*, 14(1,2,3): 174-182, 1971-1972.

Eighty-two auxiliary school MR students, divided into experimental and control classes, participated in a study of techniques of speech training directed towards student comprehension of the immediate environment. The topics "plants," "animals," and "birds" were used to develop conversational speech and fundamentals of coherent monological speech. An individual pupil was instructed to pick up an object, examine it, perform certain operations with it, and describe it. Constructive activity was employed in order to teach the Ss to organize their short speeches in a logical sequence. The overall number of sentences used in a descriptive account was found to be 1.5 to 2 times greater in the experimental group than in the control group. Under experimental conditions, a significantly greater number of children was able to enrich its vocabulary as compared with control Ss. (1 ref.) - *B. J. Grylack.*

Kiev A. M. Gorky State Pedagogical Institute
Kiev, Ukrainian S. S. R.

3259 **LOTT, GEORGE M.** Psychotherapy of the mentally retarded: Values and cautions. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 194-202.

Psychotherapy has a definite role to play in the general treatment and training of the retarded, although its contributions are still debated. Confusion in this field is attributable to the fact that slow children represent many types of defects and handicaps, and to the varieties of emotional inhibition. MRs have less effective powers of inhibition and lower tolerances of frustration, both of which can be enhanced by wise psychiatric treatment. Both those with emotional problems and those with aphasia, deafness, and speech problems can benefit from analytical forms of treatment. Despite the fact that contraindications may occasionally preclude psychiatric treatment, it can assist in the management

of certain exceptional children. (15 refs.) - *S. Glinsky.*

Mental Health Center
York, Pennsylvania

3260 **FREEMAN, ROGER D.** Drug effects on learning in children—a selective review of the past thirty years. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 257-292.

Drugs have a definite if circumscribed helping role to play in clinical work with children suffering from learning disabilities. Over the last thirty years a plethora of drugs has appeared and claims abound for their success in modifying behavior and learning. While there is genuine success, recourse to drugs must be tempered by a realization of a multiplicity of factors impinging upon their effects. The definition of learning itself is open to debate. Methodological factors include instruments for measuring improvement, the definition of improvement, administration and dosage schedule of the drugs, the placebo effect, control groups, observer bias, study designs and side effects. Recently biochemical approaches to memory and learning have shown promise. Although on balance definite progress has been made, the host of ambiguities still surrounding psychopharmacologic agents calls for great discretion in their application. Continued work in these drugs and the physiology of learning allows cautiously optimistic expectations for the future. (198 refs.) - *S. Glinsky.*

The University of British Columbia
Division of Child Psychiatry
Vancouver, Canada

3261 **KURLANDER, LEROY; & COLODNY, DOROTHY.** Panacea, palliation or poison: The psychodynamics of a controversy. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 252-256.

The decision to use medication on a disturbed child should rest on the merits of each case and the specific need of the illness. A great deal of professional provincialism and irrational anxiety clouds the medication issue. Attitudes of psychiatrists toward the use of drugs are either conscious opinions, half-conscious ones for or against drugs or unconscious responses to an agent which deeply affects a child. A rational approach dispenses with historical positions as well as with desires by psychiatrists to be doctors of the mind rather than of the body. The psychiatrist must wisely utilize all the tools at his disposal. (No refs.) - *S. Glinsky.*

University of California School of Medicine
San Diego, California

3262 **WRIGHT, MARGARET; & MENOLASCINO, FRANK J.** Nurturant nursing of mentally retarded ruminators. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 228-240.

A nursing treatment intervention program stopped rumination in young MRs. Four young institutionalized MRs were treated during an 18 month period. The treatment program began with a thorough diagnostic evaluation and consisted of an initial phase, where a relationship was established with the child, a middle phase of graduated exposures to new enjoyable experiences and a final phase of solidification of gains. The success of the treatment points up the possibility of extensive behavior modification of the more severely retarded population. A high index of suspicion on the part of nursing personnel is required to identify such syndromes. (8 refs.) - *S. Glinsky.*

Atlantic Community College
Mays Landing, New Jersey

3263 **WOODWARD, KATHERINE; JAFFE, NORMA; & BROWN, DOROTHY.** Psychiatric program for very young retarded children. In: Menolascino, Frank J. ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 204-218.

Retarded functioning without demonstrable organic basis in the very early years need not be

regarded with pessimism. A psychiatrically oriented nursery program for 26 preschool children with retarded functioning, aged 23 months to 4½ years, was conducted for varying periods of time. The children included those without demonstrable organic damage or with minimal or questionable damage. Of those who were not clearly psychotic, a high percentage showed considerably improved functioning, some even to the point of normalcy. These findings indicate that psychogenic factors frequently play a major role in the developmental lag. Treatment of both child and family at this crucial early stage is essential. (23 refs.) - *S. Glinsky.*

New York Medical College Hospitals
New York, New York

3264 LORENTZ, RICHARD J. Dyslexia. *Journal of Rehabilitation,* 37(5): 25-27, 1971.

Dyslexia, a symptom complex in which a person has difficulty in reading, writing, and spelling, is not easily diagnosed and the treatment is rather limited. It is important that the condition be detected as early in life as possible and that treatment be continued as long as necessary, usually throughout the school age or as long as the patient is making progress. Diagnosis and treatment must be carried out by physicians in cooperation with paramedical and other professional personnel in education, psychology, and sociology. A long-range remedial reading program must be available with periodic evaulation of the child's abilities even after treatment has been stopped for several years. Because of the diverse disciplines required to cope with the problem, a community approach is necessary; for the same reason, little has been done in many communities. This has opened the door for charlatans and self-styled "experts" to exploit the situation. About 10-15% of school children fail to read, write, and spell up to their proper grade level, and about 5% remain retarded in these subjects throughout life. Males are more affected than females, the ratio being from 4 to 1 to 10 to 1 depending upon the study being cited. Because of the seriousness of the problem, federal assistance is available (Learning Disabilities Act of 1969), but the amount available per child is so small that most of the financial burden for a substantial program must be borne by individual communities. There is no hard and fast approach to establishing a dyslexic program. Each community must develop its own program in line with its needs. (15 refs.) - *S. M. Rucker.*

No address

PROGRAMMATIC ASPECTS – Planning and Legislative

3265 Help for the handicapped. *Lancet*, 1(7710): 1169-1170, 1971. (Editorial)

Results of a government-sponsored survey of the impaired and handicapped in Great Britain have revealed that there are over 3 million people aged 16 years or over, currently living in private households, who have either physical, mental, or sensory impairments. The survey sample was classified according to severity of handicap, and the information collected on day-to-day living, income, housing, employment, and leisure activities was analyzed according to handicap, sex, and region. The report revealed many cases where the disabled were not getting the help to which they were entitled, frequently because they lacked information as to its availability. Glaring lapses in the provision of welfare services were revealed, problems of unsuitable housing and employment situations being particularly striking. Local health authorities, now that they have the needed information, ought immediately to begin an evaluation of their own social services patterns and generate suggestions for improvement. (1 ref.) - *N. Mize.*

3266 **MARTIN, EDWARD W.** Bureau of Education for the Handicapped commitment and program in early childhood education. *Exceptional Children*, 37(9): 661-663, 1971.

A major objective of the Bureau of Education for the Handicapped of the Department of Health, Education, and Welfare is to provide positive and constructive early education for 75% of all potentially handicapped preschool children in day care, Headstart, and early education programs by 1976. Programs funded by the Handicapped Children's Early Education Assistance Act (1968) are designed to increase the visibility and to demonstrate the effectiveness of preschool and early education programs in order to stimulate local and state efforts. The activities of the divisions of the Bureau include services to children, research, and manpower training. They are aimed at coordinating resources, materials, and ideas and at ensuring maximum involvement of national, regional, state, and local agencies in the development of appropriate preschool and early educational programing. (5 refs.) - *J. K. Wyatt.*

Department of Health, Education, and Welfare
Washington, D. C.

3267 Rapport moral (Moral report). *Nos Enfants Inadaptes*, 38 (second trimester): 3-8, 1971.

The French government has finally made plans to help to finance the employment of retarded children and to meet related demands. Although the public has demonstrated great generosity, only the government can make available the necessary credit and funds. The 1971 budget includes a loan for workshops and provides for full coverage of long illness in institutions for the retarded. In addition, the Minister of Foreign Affairs has urged the United Nations to adopt a declaration of rights for the mentally deficient. Efforts must continue towards obtaining improved living and working conditions for retarded youngsters and relief for their families. (No refs.) - *B. J. Grylack.*

3268 Transformation for the mentally handicapped? *Lancet*, 2(7714): 29-30, 1971.

The policies and recommendations of the British government's white paper on the mentally handicapped (1971) are discussed. The paper emphasizes the planned shift of custodial services from hospital to community and from central Government to local authorities, implying a significant increase in social workers and other specialized staff. Hospitals will reduce their intake of mentally handicapped patients, and no new large hospitals will be built, but outpatients, day-patients and inpatients will require new assessment and treatment facilities. Shortages of facilities, staff, and provisions for staff training at present make it questionable whether local

authorities can cope with the demands to be placed upon them. The "total transformation" of services for the mentally handicapped envisaged by the plan seems dubious in the light of limited resources and lack of any innovative ideas. (1 ref.) - *T. C. Turner.*

3269 New York State. *Annual Report of the Joint Legislative Committee on Mental and Physical Handicap.* 109 p. 1970.

The contents are divided into a discussion and analysis of the 1969 and 1970 legislative programs, recodification of the mental hygiene law, and a summary of the activities of the New York State Departments of Education, Health, Mental Hygiene, and Social Services relating to the MR and the mentally and physically handicapped. (No refs.) - *T. C. Turner.*

3270 MAC KEITH, RONALD. Where do we go from here? *Developmental Medicine and Child Neurology,* 13(4): 419-420, 1971. (Editorial)

Both researchers and clinicians in medicine must consider whether they are bringing the better parts of changes in medicine to their task of helping people. Emphasis can be placed on keeping up with advances in order to make better diagnoses of presenting symptoms in terms of anatomic or biochemical disorder. Or the physician who is impressed with current emphasis on comprehensive diagnosis of the person who has the symptom may come to accept that, while the origins of many symptoms do lie in the person affected, the culture and environment in which he lives is also of importance. Such a physician may then become concerned about the general health of children in the community, early identification of physical, mental, and psychosocial disorders, and use of epidemiological information gained from such population screenings in determining how best to deploy resources for delivery of medical care. He may feel that hospital medicine is less important than the fact that a lot is known about common disorders which is not yet put into use for the benefit of many children. Care of children with chronic neurological deficits has been neglected in the past. Help can be brought by universal periodic developmental assessment of all children in the community. This can be instituted within current resources. While research and better care must continue, perhaps we should pause to consider the best deployment and direction of research and habilitative care. (No refs.) - *K. L. Woodworth.*

Spastics International Medical Publications
London, W.1., England

3271 ELLIOTT, JAMES. Service for the mentally handicapped. *Lancet,* 2(7715): 92, 1971. (Letter)

Approximately 150,000 MR individuals have been given a white paper which promises some new, limited policies and more financial aid. Although this report represents progress, its proposals are weakened by hesitation. While it vetoes new large hospitals in favor of the small-group approach, the veto applies only for the present time. Area hospitals of up to 200 beds are recommended, but it is not specified whether these will be hospitals in traditional terms or centers for the training and socialization of SMR persons. The report should produce a number of separate better services for MRs, but it will not produce 1 united and comprehensive service without further pressure from professionals in the field of MR. (No refs.) - *B. J. Grylack.*

The Hospital Centre
London W. 1, England

3272 CAWLEY, IRIS. Transformation for the mentally handicapped. *Lancet,* 2(7719): 321-322, 1971. (Letter)

Lancet's editorial of July 3, 1971, concluded with the suggestion that the government of Great Britain has passed over an opportunity for a complete review of the country's policies towards the MR, but this was done as long ago as the 1959 Mental Health Act. Perhaps the Editor has missed an opportunity—that of insisting on the vital connection between educating the MR and placing them in appropriate jobs. There exists a gap in forward planning for the MR for with increasing automation and unemployment, finding the right jobs for them will be a demanding and specialized matter. If this is not done, the result will be a large group of expensively educated but frustrated people. (No refs.) - *K. L. Woodworth.*

Slough Society for Mentally
Handicapped Children
Englefield Green, Surrey, England

PROGRAMMATIC ASPECTS – Community

3273 **PAPPWORTH, M. H.** The Willowbrook experiments. *Lancet*, 1(7710): 1181, 1971. (Letter)

Experiments such as those recently reported at Willowbrook where MR children, as part of a research study, were deliberately infected with hepatitis, raise important ethical issues for physicians and society at large. Even though the experimenters claimed parental consent for the program, the element of coercion (parents were told that the institution was to be closed because of overcrowding, but that children who took part in the hepatitis research project could remain) must certainly nullify whatever legal validity such consent may have under U.S. law. Further, no physician is ever justified in placing the claims of society or science above his patient's welfare. In the Willowbrook case, the resultant immunization of the MR children was only the incidental byproduct of an unethical experiment and in no way justifies the means employed. (5 refs.) - *N. Mize.*

No address

3274 **PEATICK, GEORGE E.** *School Board Members and the Education of Handicapped Students: A Dissertation.* Rutgers University, New Jersey, 1970. 4p. (Abstract)

A multiple type response questionnaire was given to 610 school district Board of Education members to discover: (a) the extent to which they understood the Laws of New Jersey governing education of the handicapped; (b) information regarding school board members' views on providing or limiting programs; (c) whether selected factors were related to their knowledge and views; (d) information about the major obstacles to program provision and solutions to these problems; (e) information about board members' opinions on selected topics in the special education field. Seventy-seven percent of board members had little knowledge of the laws on the education of the handicapped. The most knowledgeable board members and the ones most likely to have positive views toward providing suitable programs for educating the handicapped were those who had a close association or friendship or were related to a handicapped person. Regionalization for special education and provision of programs for the gifted as well as the retarded were approved; paying a salary differential to teachers of special classes was not approved. Rated as most important problems were those related to finding trained personnel, adequate facilities, and financing programs. (No refs.) - *T. C. Turner.*

Rutgers University
New Brunswick, New Jersey

3275 Law & medicine–Testamentary capacity. *Journal of the American Medical Association*, 215(12): 2029-2030, 1971.

In order to make a will an individual must understand the nature and extent of his property, appreciate the nature of the claims upon him of the natural objects of his bounty, and formulate an orderly scheme of disposition. An MR person may make a will if he can comply with these criteria. In order for a will to be overthrown, it must be shown that the testator was adjudicated incompetent before his death. (No refs.) - *E. L. Rowan.*

3276 **HOLDER, ANGELA R.** Mental illness and parental rights. *Journal of the American Medical Association*, 216(3): 575-576, 1971.

In adjudicating cases of adoption and custody in which mental illness or MR is present in the parents, the paramount consideration of the courts is the best interests of the child, with parental rights of distinctly secondary importance. The common law principle that a person declared incompetent cannot give consent to a legal proceeding prevented children of such parents from being adopted; most jurisdictions therefore enacted legislation providing for

assigning of guardians for the parents who would be authorized to consent to adoptive actions for them if the condition of the parent appears to be incurable. MR, either alone or in conjunction with mental illness, may constitute grounds for termination of parental rights, but only in cases where considerable MR is evident. The ability of the parent to cope with situations involved in rearing a child is the test used to determine parental rights. (No refs.) - *J. K. Wyatt.*

American Medical Association
Chicago, Illinois 60610

3277 GRASS, CONSTANCE; & UMANSKY, RICHARD. Problems in promoting the growth of multidisciplinary diagnostic and counseling clinics for mentally retarded children in nonmetropolitan areas. *American Journal of Public Health*, 61(4): 698-710, 1971.

The findings of a survey of 4 clinics for the MR in non-metropolitan areas indicate that these clinics need to be an integrated part of a centrally regulated statewide system of services to assure quality services and viability. Locally based clinics permit the translation of diagnostic data into practical therapeutic programs for children and their families. Among the difficulties which interfered with the functioning of the various clinics were: poor administrative support and internal dissension; lack of adequate support from the community agency administering the program; unresolved interdisciplinary differences; limitations in staff time, energy, and experience; emphasis on primary diagnosis rather than follow-up; and/or inadequate funding. Two clinics are functioning fairly well; 1 has not been able consistently to offer services of sufficiently good quality; and it is too early to evaluate the success of the fourth. Effective on-the-job training is needed because clinics of this type require a higher degree of diagnostic ability and collaborative professional endeavor than is generally found among clinic personnel. (5 refs.) - *J. K. Wyatt.*

Northern California Regional
Child Development Center
Oakland, California 94609

3278 DYBWAD, GUNNAR. De nouvelles perspectives pour l'organisation de nos associations (New perspectives for the organization of our associations). *Nos Enfants Inadaptes*, 38 (second trimester): 15-17, 20-21, 1971.

The National Union of the Associations of Parents of Retarded Children is too oriented towards the past and is unprepared for the future. Rather than conceiving of mental deficiency in terms of children alone, the Union should direct its efforts towards expansion of institutions and services for all retarded individuals and towards improved programs for the lifespan of mental deficients. Sweden has set an excellent example by organizing the first conference of young mentally handicapped adults directed by these individuals themselves. Without the work accomplished by parents, local, and national associations, no progress at all would have been made towards recognition of the rights of the mentally handicapped. It is now time to reorganize efforts in order to maintain this development in the future. (No refs.) - *B. J. Grylack.*

No address

3279 ROGER, LOUIS. Le probleme angoissant de nos adultes a domine les "Journess d'Alsace" (The agonizing problem of our adults has dominated the "Days of Alsace"). *Nos Enfants Inadaptes*, 37 (first trimester): 3-5, 1971.

At a time when there are more than 200 thousand mentally handicapped in France and only approximately 4 thousand workshop positions available, the people of Strasbourg have given these individuals renewed hope through the operation of the Chateau de Bollwiller, in which mentally handicapped young people have been allowed to develop their skills and to prove their usefulness as human beings. During the conference of the National Union of the Associations of Parents of Retarded Children in Strasbourg, the question of homes for and tutelage of the handicapped was discussed. There are currently only about 500 places in homes for handicapped orphans, and all others are assigned to psychiatric hospitals. Until more homes for placement can be found, these hospitals should be modernized. It is particularly appropriate for the retarded to remain in placement homes during the work week and to return to their own families for weekends and vacations when the place of work is too far away to permit daily commuting. The

efforts of the Union will now be directed towards alerting the entire French public to the needs of the mentally handicapped. (No refs.) - *B. J. Grylack.*

No address

3280 **BROGAN, WESLEY G.; & BROWN, CECIL J.** The church and mental retardation: A guide to planning an orientation workshop for clergymen. 19 p.

The brochure prepared by the North Carolina Council on MR is designed to guide community coordinators in planning and implementing workshops for ministers and lay leaders in order to aid the churches in meeting the spiritual needs of MRs. Section One provides suggestions on persons to involve and administrative steps to follow in planning an orientation session. Section Two enumerates the main areas for consideration in an orientation session, including Sunday school classes, a weekday activity center, a sitter service, organized visiting, camping, a retreat for older adult MRs, vacation church school classes, scouting, day care services, state centers for MRs, and workshops for teachers. Not only must the church serve the retarded, but MRs can serve the church by participating in the church service, in food and child care services, in office work, and in care of the buildings and grounds. (13 refs.) - *B. J. Grylack.*

North Carolina Council on Mental Retardation
Raleigh, North Carolina 27605

3281 **BERLOGEA, OCTAVIAN.** The economics of rehabilitation as seen by the physicians in charge of county medical services. *Rehabilitation,* 77(April-June): 27-33, 1971.

The combination of the social insurance system and state medical assistance in Roumania have been decisive in the effectiveness of rehabilitation, of bringing the individual back to physical, mental, and economic usefulness. The disabled can actively participate in the production of goods for the community, which is thus relieved of the costs necessary for supporting inactive, dependent persons. In the future, the rehabilitation of the disabled will be integrated into a larger social framework more in line with the aims of social policy regarding health and economic security. (No refs.) - *T. C. Turner.*

The Institute of Medical Assessment and Rehabilitation of Working Capacity
Bucharest, Roumania

3282 **HUTCHBY, J. P.** A community occupational therapy service and its future role. *Rehabilitation,* 76(January-March): 11-14, 1971.

The Buckinghamshire community occupational therapy service is described. Covered are: present services and their future, pilot projects, administration and staffing, and recommendations for use in setting up similar institutions. (No refs.) - *T. C. Turner.*

Buckinghamshire County Council
Buckinghamshire, England

3283 **ENGLISH, R. WILLIAM; & PALLA, DARID A.** Attitudes toward a photograph of a mildly and severely mentally retarded child. *American Institute for Mental Studies Training School Bulletin,* 68(1): 55-63, 1971.

The attitudes of the general public toward the phenotype of a mildly and a severely retarded child were investigated. Ss were 80 female clerical office workers at Syracuse University who viewed 2 photographs, one of a mildly MR child, one of a severely MR child, and rated them on a scale of 20 pairs of bipolar adjectives. Opinions were far more negative towards the photograph of the severely MR child than towards that of the mildly MR child. The implications of this finding are discussed and recommendations made toward remediation of public attitudes toward the severely or obviously MR. (18 refs.) - *T. C. Turner.*

Syracuse University
Syracuse, New York

3284 **ABESON, ALAN; TRUDEAU, ELAINE; & WEINTRAUB, FRED.** Law review:

Legal opportunities and considerations for early childhood education. *Exceptional Children*, 37(9): 697-701, 1971.

Categories of state laws related to the provision of special educational services to preschool handicapped children include: legal authorization for the inclusion of these children in educational programs; administrative patterns designed to provide delivery of programs; and legal provisions for associated services. Special services for handicapped children have been provided in some states by lowering the legal age limit, requiring early education for selected categories of handicapped children, and/or making early education experiences available to exceptional children who would be irreparably damaged without them. Among the legal provisions for administrative patterns are the inclusion of preschool children in administrative policies authorized under a special education law which includes all handicapped children; the provision of special authorities for preschool programs; or the operation of programs directly from the state level within state institutions. All states have crippled children's units which provide health services, and some states provide various associated services through area or community based programs. Preschool special education legislation should provide for parent participation, early identification, and effective evaluation. (1 ref.) - *J. K. Wyatt.*

No address

3285 **DENNY, ROBERT L.** *Council on Developmental Disabilities: Annual Report, 1971.* Raleigh, North Carolina Department of Human Resources, 1971, 34p.

This 1971 Annual Report of North Carolina's Council on Developmental Disabilities comprises individual summaries by various corps of coordinators who deal with the state's developmentally disabled. The Council serves as an advisor for a state plan to receive federal funds under Public Law 91-517, which provides money for improved services and construction of facilities for those with developmental disabilities (cerebral palsy, epilepsy, MR, and other neurological disabling conditions) from birth until age 18. The report reflects the great surge of interest in these handicapped persons, their need for support, priorities for use of funds and professional personnel, and the need to enlist community support. The separate statements by the supervisors of the state's 4 regional offices and by the latter's various area coordinators cover all aspects of the Council's concerns: coordination with schools in planning programs for the handicapped, sheltered workshops, disadvantaged populations, halfway houses, workshops in speech therapy, day care, recreation, and others. (No refs.) - *B. Berman.*

Department of Human Resources
Raleigh, North Carolina

3286 **CENTERWALL, SIEGFRIED A.** The mentally retarded and the family physician: Results of a survey of Michigan doctors. *Michigan Medicine*, 70(16): 597-602, 1971.

A survey of Michigan doctors on their practice regarding MR revealed limited interest in and attention to MR. In April and May, 1968, a questionnaire regarding medical practice with respect to MR was administered by professional interviewers to a representative sample of Michigan doctors in general practice, in osteopathy, and in private pediatric practice. Only half of the general practitioners considered that they saw any MR cases at all. The average number seen by the other half was about 1 or 2 new cases per year, and about 75% of these were referred to other doctors. The pediatricians encountered an average of 10 new cases of moderate or SMR per year, and 80% of the pediatricians did the work-up and follow-up, while the remainder referred the MR cases to special clinics. Special interest in MR was limited to 1 to 2% of general practitioners and 12% of pediatricians, and only limited interest in postgraduate education was expressed by any of the doctors. Fifty per cent of general practitoners and 25% of the pediatricians believed in immediate institutionalization of mongoloid babies. Discrepancies between the number of newborn per year and new cases seen indicated much overreferring and duplication. This could be minimized by complete state supported MR diagnostic facilities and a state registry. Continuing education in MR should be directed mostly to the needs of pediatricians. (5 refs.) - *S. Glinsky.*

San Bernardino County Health Department
San Bernardino, California

3287 **SHERMAN, ANN; PAYNE, JAMES S.; & *CARRIKER, WILLIAM R.** Is Head Start

dying? *American Institute for Mental Studies Training School Bulletin*, 68(2): 113-130, 1971.

The future of the Head Start Program is discussed by reviewing the literature to date and examining the program's status along with evidence of a decline in public interest. The transfer of Head Start from the Office of Economic Opportunity to the Department of Health, Education and Welfare, while providing greater financial aid to the program's extensions, did not cause financial benefit to Head Start itself. The Westinghouse Report (first large-scale study of Head Start) stated that summer programs did not produce early cognitive and affective gains continuing through first grade. Although many social scientists and statistical experts criticized the study, the White House approved the report, while the press mostly interpreted it as pointing to Head Start's failure. Public schools were then criticized, charging that they must prepare for those who have come through the Project. Head Start tried parental involvement, teacher training, and experimental and demonstrative projects such as Sesame Street, and has been promoting health and nutritional benefits for children in the Project. Interest in Head Start is waning and it is doubtful whether it can survive in its present form. (51 refs.) - *T. C. Turner.*

*University of Virginia
Charlottesville, Virginia

3288 WALLACE, HELEN M. Children and youth projects and related comprehensive health care projects. *Clinical Pediatrics*, 10(9): 487-494, 1971.

Comprehensive health services for children and youth are currently limited. Proper coverage requires careful planning, legislation and funding of projects, and identification of the essential components. A good program ignores geographic boundaries, uses extensive community and regional planning, and is adequately funded on federal and local levels. Comprehensive care should be an integral part of care for all family members. Evaluation studies and proper standards established by experts are needed. Health care involves preventive, curative, and rehabilitative aspects. Not least are an adequate supply of manpower and innovative approaches in personnel training. (16 refs.) - *B. Berman.*

University of California
Berkeley, California

3289 SHARRARD, W.J.W. Assessment and action. *Developmental Medicine and Child Neurology*, 13(5): 561-562, 1971.

In the last 10 years there has been a marked increase in the establishment of evaluative services for physically and mentally handicapped children. The danger exists, however, that society and its doctors may satisfy their conscience and medical interests simply by continuing to make periodic assessments without sufficient reference to investigation, treatment, and comprehensive care which might benefit the child. Even when facilities and expertise exist for correcting progressive deformity or deterioration of function, the neurologically handicapped child often must wait for surgical treatment until more normal children receive it. In many teaching centers, the management of cerebral palsy and the operative measures that may benefit it are not known or not taught. Much can be done to alleviate disabilities of childhood development and neurological handicap despite lack of a cure. (No refs.) - *B. J. Grylack.*

Spastics International Medical Publications
London W. 1, England

3290 OZARIN, LUCY D.; & FELDMAN, SAUL. Implications for health service delivery: The community mental health centers amendments of 1970 (Symposium). *American Journal of Public Health*, 61(9): 1780-1784, 1971.

In 1970, the Community Mental Health Centers Act (1963) was amended again to renew the authorization and to broaden the scope of services. The primary objective of the community mental health centers program is to provide as comprehensive a range of accessible and acceptable mental health services to defined population units (75-200,000) as possible, with continuity of care and preventive measures. Another objective is to bring together the available resources in a given area into a network with defined communications links. By June 30, 1971, 452 centers had been funded, with about 300 in operation. Nearly three-fourths had general hospitals in their

organizations; other grantees included mental health clinics, mental hospitals, medical schools, and special purpose corporations, including county mental health boards. A spectacular result of the program has been the sharp decline in resident populations of State hospitals and the shift in focus from inpatient to outpatient services. The new legislation extends federal support for both staffing and construction and gives special attention to areas of urban and rural poverty. Higher levels of staffing support also are provided for services for alcoholics, narcotics users, and children. For children, federal support for a complete mental health service is possible even though a total mental health program may not exist in the community. Funds also are provided to help support consultation and education programs. (No refs.) - *S. M. Rucker.*

National Institute of Mental Health
Rockville, Maryland 20852

3291 RAGAN, CHARLOTTE E. Retarded adults in foster homes. *Rehabilitation Record,* 12(5): 37-39, 1971.

A program in Missouri known as the Sikeston-Delmo project is successfully transferring adult female MRs from institutional care into private foster homes. The immediate aim of the project, which was funded in 1970 for 3 years, is to develop small rural communities to provide family care for 60 adult female MRs. An economically depressed area in Southeast Missouri in which the Delmo Housing Corp. - a nonprofit social service agency - serves 10 communities in 5 counties was chosen for the experiment. Most families in this rural area are in need of supplemental income, and few employment opportunities exist, particularly for the women. The Sikeston Clinic, with a full-time staff of 65, provides direction of the program which fills two great needs: the patients' need for a family-like environment and the family's need for supplemental income. After appropriate ground-work in a chosen community, housewives who wish to participate are given an 8-week training course during which they receive a stipend of $20 per week which helps them to prepare their homes for their patients. After receiving their patients, they are paid $125 per month per patient and many take 2 patients. An activity center is established in the village where a varied educational and recreational program is provided from 1 to 5 o'clock each afternoon. Therapy sessions, as well as vocational training, also are provided depending upon the degree of impairment. Tax costs are reduced since the average cost per institutionalized patient is from $300 to $500 per month. (No refs.) - *S. M. Rucker.*

3292 STINE, OSCAR C.; & KELLEY, ELIZABETH B. Evaluation of a school for young mothers. *Pediatrics,* 46(4): 581-587, 1970.

Attendance at a public school program for teenage mothers resulted in significantly fewer low birth weight infants and fewer premature births than for a control group. The study group had access to prenatal care, school lunch, supplemental milk, counseling, and nutrition and health education. The study and control groups totaled 448 births to mothers 14, 15, and 16 years old. Eleven point six per cent of the study group and 23.7% of the controls had low birth weight infants, and 21.4% of the study group versus 34.4% of the controls had premature births. One infant died in the study group and 8 in the control group. These results confirm the value of such programs. (6 refs.) - *S. Glinsky.*

University of Maryland
Baltimore, Maryland

PROGRAMMATIC ASPECTS — Residential

3293 Buildings for the mentally handicapped. *Lancet,* 1(7711): 1249, 1971.

Britain's hospitals for the MR commonly share problems of chronic overcrowding and understaffing. Recommendations from a recently appointed study group of architects and allied professionals in the MR field are deficient to the extent that they deal only with overcrowding and leave the second problem undiscussed. Since the two are interrelated, alleviating overcrowding by increasing patient/floor space ratios, as recom-

mended, will lower still further the already abysmal patient/staff ratios. Before any new suggestions are implemented, further critical study must be given to the cost and care advantages of the traditional hospital-based system, on which the committee's report is grounded, as compared to the newer ideas involving community-based supervision of the MR. (1 ref.) - *N. Mize.*

3294 **HEATH, P. J.** A long-stay therapeutic community. *Lancet*, 1(7711): 1231-1232, 1971.

A therapeutic community for a mixed group of long-stay patients – geriatric, psychogeriatric, MR, mentally ill, chronically sick, and drug addicts – is advocated as a needed replacement for the large mental hospitals which have traditionally housed these patients in Britain. The suggested reorganization is similar to the balanced hospital community which has been previously proposed by others concerned with the problem of longterm care. Emphasis would be on establishing strong and numerous links with the surrounding community, a feature which should greatly facilitate the necessary dual process of education for both patients and the larger society within which they live. Groups of patients suitable for vocational work, such as the high grade MR, drug addicts, and young psychiatric patients, would hold jobs either in the nearby town or within the therapeutic community's own sheltered workshops. Every effort would be made to provide all needed medical services, as well as those additional facilities, such as libraries, hairdressers, shops, etc., which contribute to the patients' social integration. (3 refs.) - *N. Mize.*

Sheffield Regional Hospital Board
Sheffield, England

3295 Mental retardation. *Children*, 18(3): 113, 1971.

Harold Decker had designed a system for providing comprehensive health care in resident institutions for the MR. Provisions are made for appraising each resident's present clinical status and level of functioning, establishing individual health and rehabilitation goals and a plan of care for each patient, following up to see that the plan is carried out, and re-evaluating the plan periodically and making needed revisions. (No refs.) - *T. C. Turner.*

University of Arkansas Medical Center
Little Rock, Arkansas 72201

3296 **HERBERG, EDWARD N.** Comparative treatment technologies and health care patterns in institutions for mentally retarded. *American Journal of Public Health*, 61(5): 988-999, 1971.

A survey of 5 public institutions for the MR analyzed the deleterious and desirable effects of their technological structures on the patterns of health care rendered. The institutions were widely diversified in regard to size, geographic location, character of resident population, treatment orientations, financial resources, staffing patterns, and formal organization. The only institutions that maintained equity in service for new and long-term residents were those which were educationally oriented. These institutions also provided a more comprehensive spectrum of health care as well as greater numbers of educational, social, and developmental services. Medically oriented and educationally oriented institutions provided equal medical services. Institutions with a medical orientation had ad hoc services for disease crises and appeared unsuitable for the provision of comprehensive habilitative care. (10 refs.) - *J. K. Wyatt.*

University of Michigan
Ann Arbor, Michigan 48103

3297 Une reussite de Marseille, le foyer du Centre des Pins (A happy outcome in Marseille, the Centre des Pins home). *Nos Enfants Inadaptes*, 37 (first trimester): 17-19, 1971.

In 1967 the Centre des Pins home in Marseille, France, opened its doors to SMR individuals with IQs ranging from 30 to 50. The institution can accommodate 27 residents and gives priority to orphans or to persons otherwise deprived of a family environment. Personnel are selected carefully for their practical skills, understanding, and compassion. The Centre des Pins represents a miniature society in which educators and MRs

are tightly bound by a common way of life. Numerous excursions are planned in order to give the residents contact with the outside world. The notions of exchange and communication, which are indispensable to the progress of the retarded, are underlying principles at the Centre and contribute to the great degree of solidarity experienced by the residents and staff. (No refs.) - *B. J. Grylack.*

3298 **KLEIN, STANLEY D.; & ABRAMS, SUSAN L.** Public housing for the handicapped? *Journal of Rehabilitation*, 37(2): 20-21, 1971.

The development of specialized housing for the physically and mentally handicapped within housing projects for the nonhandicapped members of the community is proposed. The construction of such housing presents few architectural problems, and physically and/or mentally handicapped persons do not need constant psychological or medical attention. Three prototype housing settings are mentioned. It is thought that integration of the physically and/or mentally handicapped into public housing programs is entirely feasible. (No refs.) - *T. C. Turner.*

University of Massachusetts
Boston, Massachusetts

3299 **GIBSON, J.** Transformation for the mentally handicapped. *Lancet*, 2(7720): 383, 1971. (Letter)

Although the complaint has been made that the mentally handicapped who cannot live in the community are doomed to institutionalization in hospitals, hospital personnel themselves have built a new way of life for these individuals. In addition to providing all the necessary medical and nursing services, a hospital in Surrey, England, has educational, recreational, and occupational facilities for its MR patients. These activities are probably representative of services provided for MRs in similar hospitals. (No. refs.) - *B. J. J. Grylack.*

St. Lawrence's Hospital
Caterham, Surrey CR3 5YA, England

3300 **CORTAZZO, ARNOLD D.; & ALLEN, ROBERT M.** The divisional approach to achieving hospital improvement goals. *American Institute for Mental Studies Training School Bulletin*, 67(4): 235-243, 1971.

A 3-year Hospital Improvement Project grant awarded by HEW's Social and Rehabilitation Service to Sunland State Institution in Miami, Florida, is being devoted to the multifaceted needs of the retarded, to identify the institution's problems, and to explain its rationale for change. The institution's evaluation, in which residents are serving as their own controls, seeks to reveal differences in pre- and post-tests on various measures. Qualitative evaluation of data include records of patients' and guardians' reactions over a period of time. Employee reactions and turnover data will be analyzed. An advisory team is evaluating management of divisional areas. The new approach to improvement is "divisional" or "horizontal," as opposed to the "traditional" or "vertical" method more usually employed. (11 refs.) - *B. Berman.*

Sunland Training Center
Miami, Florida

3301 **CORTAZZO, ARNOLD D.; BRADTKE, LOUISE M.; KIRKPATRICK, WILLIAM J., JR.; & ROSENBLATT, KATHERINE P.** Innovations to improve care in an institution for the mentally retarded. *Children*, 18(4): 149-154, 1971.

At the Sunland Training Center in Miami, a state institution for the MR, the department-centered traditional organizational model was changed to a horizontal structure for providing services. Four program divisions, each staffed by a director and an interdisciplinary team, now focus on resident-oriented goals: development and training, independent living, education and training, and vocational rehabilitation. Another innovation is the BKR Experimental Project (named after its originators) for preschool SMR to PMR children. Forty-five of the 60 children originally involved (1968-69) had been rejected from educational activities due to behavioral or intellectual deficits. Individual attention is emphasized and specific behavioral objectives are established for each child. To obtain the personnel needed, nonprofessional persons who had been unsuccessful in

their other roles on the staff were trained as teacher's aides to work with 2-4 children under a teacher's supervision. Of the original 60 children, 35 have developed to a level enabling enrollment in Sunland's formal educational program. The BKR Project has been expanded to help SMR children living at home by training their parents in behavior management techniques. (2 refs.) - *K. L. Woodworth.*

Sunland Training Center
Miami, Florida

3302 **LAWLOR, T.** Care of the mentally handicapped. *Lancet,* 2(7722): 494, 1971. (Letter)

Rising unemployment will create great joblessness among the mentally handicapped in Britain, and those responsible for services to these handicapped should prepare for such emergencies. Subnormality hospitals and staff should not be disbanded, since they best understand the needs of the handicapped and can perform services for the unemployed. (No refs.) - *B. Berman.*

Normansfield, Teddington
Middlesex, England

3303 Services for the mentally handicapped. *Lancet,* 2(7717): 208, 1971.

British Parliamentary debate on services for the mentally handicapped urges Government commitment to improved community care and facilities, but, at the same time, opposes wholesale abolition of hospitals for the mentally handicapped, since community and family care represent only an ideal. The mentally ill might be cared for in units or annexes of general hospitals, and specialist hospitals should be phased out. Patients should be placed in the appropriate residential unit, either in home or hospital. Government objective is to provide first-class detection and assessment of mental handicaps, and a full range of support, educational, and training services. (No refs.) - *B. Berman.*

3304 **NEWCOMBE, J.** Care of the mentally handicapped. *Lancet,* 2(7718): 264, 1971. (Letter)

It has been suggested by certain people that hospitals for MRs in England should be closed, that many MRs could live in small groups, that those needing hospital care could be placed in special wards in new general hospitals. There are many disadvantages, and this view is an oversimplification of the problem. Children enjoy the space and the wide range of activities in the present hospitals which would not be possible "in the cramped surroundings of the prefabricated hostels in the middle of busy towns." Nurses in general hospitals are not interested in MRs, while enormous knowhow has been built up in hospitals for MRs by people interested exclusively in this discipline. Mass transfer of MRs from hospitals to small town units would be of no benefit to the MRs. (No refs.) - *S. M. Rucker.*

Claypenny Hospital
Easingwold, Yorkshire Y06 3PR England

3305 **LORING, JAMES.** Services for the mentally handicapped. *Lancet,* 2(7716): 161, 1971. (Letter)

The white paper, *Better Services for the Mentally Handicapped,* is essentially a political document. About two thirds is devoted to uncontroversial statements about MRs and the remainder to a statement of intent on the part of the British government. A very large number of patients in hospitals for MRs do not require medical nursing care and with suitable rehabilitation could be absorbed into the community. In other than exceptional cases, children should not be kept in long-stay institutions. The white paper suggestion that nonambulant children require hospital attention is not borne out by experience. Many nonambulant MRs, with physiotherapy, can become partially ambulant. The most serious consequence of keeping the long-stay hospitals are the huge cost and the waste of medical and nursing skills which are in such short supply. (No refs.) - *S. M. Rucker.*

Spastics Society
London W1N 4EQ, England

3306 **PILKINGTON, T. L.** Staffing of mental subnormality hospitals. *Lancet,* 2(7716): 161, 1971. (Letter).

The concept of MR as a specialty in its own right is challenged by a report of the Scottish Home and Health Department on staffing of mental deficiency hospitals, which proposes a system of joint appointments. About half the total mental deficiency consultants in Scotland now hold joint appointments, and a similar system is appearing in England and Wales. Perhaps the person who provides continuity of responsibility for the MR child or adult should not be either psychiatrically or medically qualified. (1 ref.) - *S. M. Rucker.*

No address

3307 **MADDOCK, ERNEST.** Do cost variations reflect need variations...in the education and training of the institutionalized retardate? *American Institute for Mental Studies Training School Bulletin*, 68(2): 69-76, 1971.

Administrative policies affecting the educational programs of the institutionalized MR are discussed. Educating the MR in state school and mental hospitals represents an enormous financial expenditure, but a program of such magnitude has failed to spawn significant research effort; as a result there is a dearth of available reliable evidence regarding extent, degree, and effectiveness of administrative policies governing the implementation of programs in state operated schools and hospitals for the MR. It is urgent that each MR person have an opportunity to develop his/her potential, since MR children rapidly become MR adults and what happens in the intervening years can make a difference in their personal life and the future economy of the district, state, and nation. Although spending money does not assure quality in instruction and training, quality is not possible without expenditure. An institution cannot maintain the status quo and keep providing its residents fair opportunity to develop. As an illustrative case, information regarding the fiscal affairs of Pennsylvania's state school and hospitals is given in 4 tables; cost-varied and reflected need variations in the 9 institutions are cited. But to assume that a need for structured learning experiences does not exist because that need is not expressed on account ledgers in terms of expenditure is erroneous. Special education techniques are predominantly structured about practical, concrete learning experiences, which require concrete supplies. The question is asked: Can an institution which spends $.13 per year per pupil for textbooks really be meeting the needs of its individuals? (6 refs.) - *T. C. Turner.*

American Institute for Mental Studies
Vineland, New Jersey

3308 Care of violent patients. *Lancet*, 2(7715): 112, 1971.

A leaflet has been published under the auspices of the National Association for Mental Health which contains guidelines for the care of patients exhibiting violent behavior. The leaflet emphasizes that physical methods of control of these patients are necessary only when all other methods have failed. If segregation is unavoidable, it should be used only as long as is necessary to calm the patient. In all emergencies, the use of restraint should be therapeutic, never punitive. Electroconvulsant therapy (E.C.T.) should not be employed to subdue excitement or aggression, and drugs for this purpose should be used very cautiously. Whatever rules governing physical methods of control are adopted in a hospital, they should be consistent with a policy which has been approved at all levels of management and which is known to and acceptable to all staff. It is rarely necessary to resort to physical methods of control if staff morale is high. (1 ref.) - *B. J. Grylack.*

3309 **ROSEN, M.; DIGGORY, J. C.; FLOOR, LUCRETIA; & NOWAKIWSKA, MYRA.** Self-evaluation, expectancy and performance in the mentally subnormal. *Journal of Mental Deficiency Research*, 15(Part 2): 81-95, 1971.

The effects of institutionalization on goal-setting and expectancy of ultimate success on a manual assembly task were tested with institutionalized and noninstitutionalized MR students, who were matched for CA and IQ, and with normal MA and CA controls. Ss in each of the 4 groups performed the same task under both the level of aspiration and performance prediction motivational conditions. The relation between measures of self-evaluation and levels of goal-setting and expectancy of success was found to be highest in institutionalized subnormals and normal CA controls and lowest in noninstitutionalized subnormals and normal MA controls. Institutionalized and noninstitutionalized retardates showed more striking similarities than differences, and

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both groups differed significantly from normals with the same CA. It is possible that negative effects of institutionalization are limited to those which occur early in life and are reduced in severity in later years. The higher correlation of expectancy and performance with MA than with CA or IQ for all Ss suggests that performance decrements among subnormals reflect developmental lags rather than qualitative differences. (22 refs.) - *B. J. Grylack.*

Elwyn Institute
Elwyn, Pennsylvania 19063

3310 **ZUBRILIN, IU. K.** Collaborative work of pedagogues and physicians at institutions for severely mentally retarded children. *Soviet Education,* 14(1,2,3): 82-89, 1971-1972.

In the light of the availability of psychotropic agents for reducing or even eliminating excessive excitation or inhibition as well as for stimulating the development of SMR children within certain limits, the joint medical and pedagogical approach has become predominant in a large number of institutions for these children. Collaboration between physicians and teachers has had a favorable effect on the training, upbringing, and medical care of SMR children at the Murygino Children's Boarding Home of the Kirov Regional Department of Social Security. Teachers and physicians cooperate in admitting and assigning children to the various departments, in making the nosology and functional diagnosis of their diseases as accurate as possible, in prescribing therapy and observing its effects, and in preparing and attending medicopedagogical conferences. As a result of well-organized workshop training and beneficial treatment, it has been possible to secure employment for half of the children with the least damage to the central nervous system and with relatively intact learning capacities who have been released from the home in the past 3 years. This and similar experiences have demonstrated that the occupational and social adaptation of SMR children can be furthered when special appropriate conditions are created. (No refs.) - *B. J. Grylack.*

No address

3311 **D'IACHKOV, A. I.** Entries from the Soviet Pedagogical Encyclopedia: Defectological Institute, defectological education, and defectology. *Soviet Education,* 14(1,2,3): 194-206, 1971-1972.

The Institute of Defectology is a scientific research institution which studies particulars in the development of abnormal children, ways of overcoming abnormal development, the correction of and compensation for physical and mental defects, and elaboration of systems for the upbringing, education, training, and gainful employment of persons with physical and mental handicaps. Professional training is given to teachers who work with abnormal children at all age levels in various types of special institutions, including those for children with aural and visual impairment as well as MR. Based upon the general laws of development of anomalous children and the principles of their upbringing and training, each scientific disciple relating to defectology reveals typological differences in the development of children afflicted with a given defect. (2 refs.) - *B. J. Grylack.*

No address

PROGRAMMATIC ASPECTS – Recreational

3312 **LEVY, J.** An intrinsic-extrinsic motivational framework for therapeutic recreation. *Therapeutic Recreation Journal,* 5(1): 32-38, 1971.

A theoretical framework is presented to provide some guidelines for a workable model in therapeutic recreation. Recreation's key role in promoting mental health is in its potential for promoting therapeutic experience contributing to the satisfaction of intrinsic needs. The therapeutic recreator can help a retarded person by providing a social milieu free of compulsion so that he can be exposed to self-actualization experiences and can enhance his sense of dignity and personal worth. Recreation can be a socially therapeutic medium for the person who constantly avoids any situations which would be

detrimental to his frail self-image. The goal is to reorient such a person's motivation from extrinsic to intrinsic reward in order to encourage reliance on the patient's own psychological reserve in bringing about satisfaction and personal responsibility. (4 refs.) - *T. C. Turner.*

University of Illinois
Children's Research Center
Urbana, Illinois

3313 **MITCHELL, HELEN J.** A community recreation program for the mentally retarded. *Therapeutic Recreation Journal*, 5(1): 3-8, 1971.

Municipal recreation programming in relation to the development of the Washington, D. C., Department of Recreation's programs for the retarded is discussed as one method for developing a community program in an urban setting. Fifteen separate programs in 9 recreation department regions were set up to serve over 300 retarded, after a survey was made to discover how many retarded there were within the confines of the recreation regions and what kind of programs they most wanted. Other existing year-round programs are also briefly mentioned. (No refs.) - *T. C. Turner.*

National Recreation and Park Association
Washington, D. C. 20006

3314 **CONNOLLY, AUSTIN J.** Classroom forum. *Focus on Exceptional Children*, 3(1): 12, 1971.

Techniques are discussed for planning a meaningful field trip for a junior high class of EMR persons. Included are: listing things to be seen and questions the class would like answered; collecting information before the trip; discussing ways to document the trip; establishing safety and behavioral standards. The teacher must ask himself if the projected trip will be interesting, attention-holding, and contain learning value. He should contact personnel at the site of the field trip and, if possible, take the trip in advance. Management problems can be minimized by keeping to a definite routine. (No refs.) - *T. C. Turner.*

University of Missouri
Columbia, Missouri

3315 **COTTER, VANCE W.; & SPRADLIN, JOSEPH E.** A nonverbal technique for studying music preference. *Journal of Experimental Child Psychology*, 11(3): 357-365, 1971.

A nonverbal technique for studying music preference was studied in 66 retarded children and adolescents (CA 9.5 to 20.6 years) by evaluating the preference for music as compared to silence and white noise and the preferences for 2 types of music (Motown Sound and Doc Severinsen). In judging music versus silence, all Ss depressed keys resulting in music for more than half of the total time of the sessions. Music was also preferred to white noise. Fifty-nine of 66 Ss preferred Motown Sound more frequently than Doc Severinsen. The data indicate that the nonverbal technique used was reliable for evaluating listening behavior for certain types of music and other types of auditory stimuli. (10 refs.) - *B. J. Grylack.*

University of Kansas
Lawrence, Kansas

3316 **KIDD, FRANCIS T.** Summer day camping for the mentally retarded. *Therapeutic Recreation Journal*, 5(2): 91-92, 1971.

In 1969, the D.C. Department of Recreation sponsored a summer day camp for MR of all ages. Staffed by counselors trained in camp management and recreational activities, the program accepted MR with secondary disorders such as behavior problems, aphasia, and partial-sightedness. The morning program offered individual or group activities such as crafts, nature lore, ball games, and hikes. After a free lunch provided by the Office of Youth Opportunity and a rest period, additional activities—games, creative drama, swimming, clay modeling, and painting—were offered. Those participating felt that the program was a success, the counselors finding work with the MR rewarding and the MR expressing a desire to return next year. (No refs.) - *T. C. Turner.*

3317 **HEIMARK, ROBERTA; & McKINNON, RACHEL.** Leisure preferences of mentally retarded graduates of a residential training program. *Therapeutic Recreation Journal*, 5(2): 67-68, 93-94, 1971.

A study was made of 136 MR Ss living in an institution to determine how they used their leisure time. The 5 most popular activities, in descending order, were watching TV, community recreation programs for MR, movies, household tasks, and walks. Most activities were home-centered. There was also considerable interest in sports. (No refs.) - *T. C. Turner.*

Agnews State Hospital
San Jose, California

3318 **McCORMICK, JAMES.** The role of perceptual-motor training in therapeutic recreation programs for the mentally retarded. *Therapeutic Recreation Journal,* 5(2): 63-66, 1971.

Major findings on the role of perceptual-motor training in therapeutic programs for the MR are delineated. Perceptual-motor training is defined, its necessity outlined, and the manner in which to incorporate perceptual-motor activities into a therapeutic training program is discussed. It has been found that it is usually possible to equalize perceptual-motor disabilities through a progressively structured program. (6 refs.) - *T. C. Turner.*

University of Rhode Island
Kingston, Rhode Island

3319 **HAYES, GENE A.** Activity analysis: Finger painting for the mentally retarded. *Therapeutic Recreation Journal,* 5(3): 133-138, 1971.

Finger painting was selected as a recreational activity for MRs, both for its value as an art medium and for its therapeutic benefits. Since MRs have learning, motor, and communication disabilities, the finger painting process, which can be perceived directly, allows the child to successfully solve a learning situation. Though social interaction is limited, it could be used to slow down the hyperactive and to involve the withdrawn in activity in the presence of other individuals. It may be used in guiding the child to gain emotional satisfaction and social approval and to develop sensory motor skills, coordination of hand-eye movements, cognition, evaluation, and convergent production. The activity also offers great opportunity for study of MRs by therapists. Observation of the entire activity, the physical manifestations and verbal interactions of the child, and the painting itself, can be important in making differential diagnosis in brain damage, neurological impairment, MR, and emotional disturbances. (7 refs.) - *R. Seevaratnam.*

Texas Woman's University
Denton, Texas

FAMILY

3320 **WOODMANSEY, A. C.** Parent guidance. *Developmental Medicine and Child Neurology*, 13(9):243-244, 1971.

The physician must use psychotherapeutic techniques in interviewing the parents of an MR child. He must elicit diagnostically useful information and also relieve parental stress. The parents need help in solving the problems as they see them and in understanding their child's difficulty. The physician must not be emotionally detached but must empathize with parents without abrogating his medical responsibility to pursue the best treatment program. (10 refs.) - *E. L. Rowan.*

The Children's Hospital
Sheffield, S10 5DN, England

3321 **KAUFFMAN, JAMES M.; WEAVER, S. JOSEPH; & WEAVER, ANN.** Age and intelligence as correlates of perceived family relationships of underachievers. *Psychological Reports*, 28(2): 522, 1971.

The Lorge-Thorndike and Family Relations tests were administered to 41 boys and 5 girls (CA 8 to 16 yrs) referred to a remedial reading program in order to investigate the feelings of the Ss towards members of their families and their perception of the reciprocal regard for them by their families. Correlations between IQ and number of negative feelings concerning family relationships were not significant. However, more intelligent Ss tended to express greater positive feelings involving their fathers ($r=.402$, $p<.01$) and themselves ($r=.396$, $p<.01$) than less intelligent Ss. The data suggest that the perceived family relationships of more intelligent children are characterized by greater feelings of self-worth, social acceptability, and identification with the like-sexed parent and that older children tend to denigrate certain family relationships, possibly in deference to peer relationships. (1 ref.) - *B. J. Grylack.*

University of Virginia
Charlottesville, Virginia

3322 **FISCHHOFF, JOSEPH; WHITTEN, CHARLES F.; & PETTIT, MARVIN G.** A psychiatric study of mothers of infants with growth failure secondary to maternal deprivation. *Journal of Pediatrics*, 79(2): 209-215, 1971.

To ascertain whether mothers of infants with growth failure secondary to maternal deprivation evidenced character disorders, 12 such mothers each underwent 2 extensive psychiatric interviews. Infants were 3-24 months old, of normal birth weight and at least 9 months gestation, below the third percentile on the Stuart growth grids in height and weight, without organic cause for failure to thrive, and had little handling by mother. Ten mothers had enough of the following characteristics to be diagnosed as character disorders: disturbed early childhood, poor day-to-day performance, indications of fairly severe psychopathology, desire for anaclitic relationship with intense need to be cared for, literal and concrete thought patterns with limited capacity for abstraction or thinking ahead, use of denial, isolation and projection as mechanisms of defense, and tendency toward action or acting out. The other 2 mothers were classified as psychoneurotic. The features of character disorders are conducive to inadequate mothering. A problem-solving, psychotherapeutic approach will have little success. Intervention involving action and someone for the mother to lean on and show her how to care for her infant is needed. (8 refs.) - *K. L. Woodworth.*

Children's Hospital of Michigan
Detroit, Michigan 48202

3323 **WALKER, J. H.; THOMAS, M.; & RUSSELL, I. T.** Spina bifida—and the parents. *Developmental Medicine and Child Neurology*, 13(4): 462-476, 1971.

The social and emotional effects on the family of the birth and care of an infant with spina bifida cystica were studied by interviewing parents of 107 spina bifida children (48 males, 59 females, mean CA 16.5 months). Data gathered confirmed

some known epidemiological factors: lower incidence in upper 2 of 5 social class categories, high incidence among relatives, and a higher than average maternal age (17 mothers over 35 years). Dissatisfaction with the method of being told of the infant's disability was expressed by 50% of the fathers interviewed and 60% of the mothers. Very few had any real early appreciation of the extent of the possible handicap. Mother-infant separation in the hospital produced great anxiety before the mother's first visit to the child. When taking the baby home, most parents (75) said that they received no special instructions or advice about management. Support by health visitors and general practitioners varied widely; few mothers spoke warmly of the help received from health visitors. Many parents found joint clinics for providing comprehensive care a disturbing and anxiety-producing experience, with the anxiety prohibiting effective communication. Only 52 mothers fully understood the implications of further pregnancy; 43 parents felt marital relationships had deteriorated; increased tension at home was felt by 77 mothers and 56 fathers; 73 mothers felt isolated; 79 mothers were worried or depressed; only 26% of the families received help from neighbors. (11 refs.) - *K. L. Woodworth.*

The Medical School
Newcastle upon Tyne, NE1 7RU, England

3324 **FREESTON, B. M.** An enquiry into the effect of a spina bifida child upon family life. *Developmental Medicine and Child Neurology*, 13(4): 456-461, 1971.

Parents of 2 cohorts of spina bifida children (47 1-year-olds, 38 4-year-olds) were interviewed to determine the effect of a spina bifida child upon family life and to assess the adequacy of existing social services. Seven areas of family stress were identified: shock and grief at birth, as well as lack of comprehension of any explanations given; separation and visiting problems at first hospitalization; visiting difficulties (time and expense) at subsequent hospitalizations, and emotional strain on the child of the hospitalization; frequent outpatient visits; genetic counseling and family planning (although nearly all parents remembered correctly genetic information about risks involved in subsequent pregnancies, less than 1/2 of 1-year-olds' parents had sought family planning advice, and only 1 in 5 of 4-year-olds' parents had taken it); concern about whether severely handicapped children should be encouraged to live; and financial stress. A health visitor called regularly in about 1/2 the cases but could not offer much advice specific to spina bifida. The same was true of family doctors. While parents' associations could reduce parents' sense of isolation, over 1/2 the families had not joined a local group. (2 refs.) - *K. L. Woodworth.*

The University Department of Child Health
Sheffield S10 2TH, England

3325 **WOLFENSBERGER, WOLF; & KURTZ, RICHARD A.** Measurement of parents' perceptions of their children's development. *Genetic Psychology Monographs*, 83: 3-92, 1971.

Parents of EMR children are realistic in assessing their children's current MR level but unrealistic regarding future development. A program for family management in MR evolved 2 parental realism assessment techniques, which assessed parents' concurrent realism about their child's behavioral attainments and parental predictive realism about future development of their child. In a study of 190 parents of 117 EMRs seen at an evaluation center parents were quite realistic about their children's concurrent retardation but overoptimistic about improvement, especially in academic achievement. Protestant parents, parents of a high socioeconomic level, and parents with a less retarded child were more realistic than Catholic parents, parents of low socioeconomic level, and parents with SMR children. The study should be replicated with tighter controls and a larger sample. (97 refs.) - *S. Glinsky.*

University of Nebraska College of Medicine
Omaha, Nebraska 68105

3326 **JEFFREE, D. M.; & CASHDAN, ASHER.** The home background of the severely subnormal child: A second study. *British Journal of Medical Psychology*, 44: 27-33, 1971.

SMR children living at home show a relative deficit on both stimulation and affection measures. In a second study of the home background of SMRs, a group of 28 SMRs were matched individually with normal children for

MA, sex, and socioeconomic background. The SMRs lived at home but attended day training centers. On parental interview assessment the SMRs and normals differed significantly both overall and in the affection area, but the stimulation difference was not statistically significant. On 4 objective tests of experiences there were clear differences overall as well as on 3 of them taken individually. (20 refs.) - *S. Glinsky.*

Manchester University
Manchester, England

3327 **ARASTEH, JOSEPHINE D.** Parenthood: Some antecedents and consequences: A preliminary survey of the mental health literature. *Journal of Genetic Psychology,* 118 (Second half): 179-202, 1971.

Surveys of mental-health literature on parenthood reveal a need for more rigorous methodology and experimental design in this field. This survey covers both antecedents (sexual attitudes, family planning, etc.) and consequences (family size or density, ordinal position, and sex of child) of parenthood. Importance of early sex education and contraceptive information is stressed, and differing sex norms are compared. In much family-planning research, "unwanted" pregnancies and therapeutic abortions are a major concern. Too few studies have dealt with the psychological aspects of sterilization: about 100,000 men and women in the U.S. annually undergo voluntary sterilization. There is now evidence for better family-planning counseling in psychiatric clinics. Of crucial importance are family-size preferences and the effects of number of children on parents and on the child. Interest in ordinal position (birth order) of a child goes back to Galton. Ordinal position at birth is related to significant parameters; it seems the youngest member of large families is overrepresented in mental hospitals. (104 refs.) - *B. Berman.*

National Institute of Mental Health
Bethesda, Maryland

3328 **MENOLASCINO, FRANK J.** Parents of the mentally retarded: An operational approach to diagnosis and management. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 160-171.

An operational reevaluation of the three main crises afflicting MR parents can lead to a clearer diagnosis and more effective treatment. The first crisis is the shock of initial diagnosis, for which the parents need emotional support, specific diagnostic information, and guidance in parent-professional treatment plans. The second crisis is that of personal values which clash with the acceptance of the child. Treatment is primarily extensive family counseling and formal individual psychotherapy. The third crisis is the reality crisis, which can be alleviated by affording practical advice in home management, day care, and possible residential placement for the MR. MR must remain a family matter with specific guidance from professionals at times of crisis. (28 refs.) - *S. Glinsky.*

University of Nebraska College of Medicine
Omaha, Nebraska

3329 **CHESS, STELLA.** Treatment of the emotional problems of the retarded child and of the family. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 179-193.

The prevention and treatment of emotional problems of the retarded child and his family require an orientation toward using the parent as a colleague. Present day psychiatric concern with childhood MR originated historically with psychiatric treatment of childhood MR considered not to have an organic base. Modern child psychiatry recognizes five distinct areas of MR, including MR with no behavior disorder, with behavior disorder due to cerebral dysfunction, with reactive behavior disorder, with neurotic behavior disorder, and with psychosis. Each type requires a different treatment approach. The child's temperamental organization is an important variable. Finally, parental guidance is seen as a specific therapeutic tool. (No refs.) - *S. Glinsky.*

New York University School of Medicine
New York, New York

3330 **MOWATT, MARIAN.** Emotional conflicts of handicapped young adults and their mothers. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 153-159.

Separate group discussions of emotional conflicts in parents and their teenage MR children clarified problems and alleviated some of the conflicts. A series of eight biweekly group discussions were held for mothers of MRs, and six weekly meetings for their MR children, who suffered predominantly from cerebral palsy. Problem areas explored included MR independence from the family, expression of sexual interest, and social communication. The mothers apparently harbored more unresolved feelings of conflict, which appeared to be more crippling emotionally than the young people's physical handicaps. Group participation of both parents and MR children simultaneously can perhaps resolve more conflicts and lead to more independent functioning for both generations. (3 refs.) - *S. Glinsky.*

Clinical Psychology
Areis Building
Seattle, Washington

3331 **SOLNIT, ALBERT; & STARK, MARY.** Mourning and the birth of a defective child. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 114-125.

Knowledge of the theory of the mourning process is essential in order to understand the depressed reactions of mothers to the birth of an MR child. A mutuality exists between theory and clinical work, which affords the pediatrician a general theory of human behavior and sharpens the analyst's theoretical deductions. Investigation of the grief reaction in mothers who have given birth to defective children has shown that the physician's awareness of the mourning process enabled him to provide effective therapeutic help to the mother. In order to facilitate the work of the mourning process the mother requires physical rest, an opportunity to reflect on her thoughts and feelings regarding the wished-for child, a realistic interpretation of the unwanted child by doctors and nurses, and an active role in planning and caring for the newborn child. (28 refs.) - *S. Glinsky.*

Yale University School of Medicine
New Haven, Connecticut

3332 **SOLOMONS, GERALD.** What do you tell the parents of a retarded child? Some personal experiences and reflections. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 144-152.

Advice and help given to parents with an MR child should be realistic, understandable, and keyed to parental, not professional, responsibility for the child. Steps to be taken by the physician in counseling and testing include listening carefully to the parents, reviewing the child's history, interpreting both positive and negative findings, and giving as clear a diagnosis and prognosis as possible. Institutionalization and special education are for the parents to decide. Treatment plans must be carefully detailed, the frequency of return appointments watched closely, and all major decisions left in the hands of the parents. (6 refs.) - *S. Glinsky.*

University of Iowa College of Medicine
Iowa City, Iowa

3333 **BEGAB, MICHAEL J.** Mental retardation and family stress. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 128-142.

The success of the MR depends largely upon how well the family can handle the stress the MR's presence creates. MR is primarily a family problem and an assessment of familial stress and treatment plans requires careful individualization of each situation. Stress factors upon family life include psychological and emotional reactions with their effect upon family stability, care and management problems, educational needs in guidance and training, and planning for future

care. Appropriate solutions to each family situation can be undertaken only when all stress and reaction factors in both the family and community are recognized. (8 refs.) - *S. Glinsky.*

National Institute of Child Health
and Human Development
National Institute of Health
Bethesda, Maryland

PERSONNEL

3334 **BLUM, EVELYN R.** Conversations in training and research. Interview with Jasper Harvey: Development of a staff training prototype for early childhood centers. *Exceptional Children*, 37(9): 670, 672-675, 1971.

The major functions of early childhood staff training programs are the development of a flexible staff training prototype based on a team concept, the provision of direct services to training facilitators in 41 centers, and the development of a preschool demonstration training site. Among the simulation techniques used in the model classroom are microteaching, role playing, team teaching, and observation. The role of consultant is under examination, and a model to evaluate the effects of consultants is being developed. (No refs.) - *J. K. Wyatt.*

University of Texas
Austin, Texas

3335 **JOLLY, HUGH.** The role of the paediatrician in the care of the mentally handicapped. *Developmental Medicine and Child Neurology*, 13(9): 240-242, 1971.

The pediatrician can play an important role in both recognizing and caring for the MR child. The proper assessment of MR may require long-term observation of the child in the company of normal children. Caring for a few long-term MR patients on a general ward allows the physician to work with the parents in a more normal atmosphere and increases the chances of rehabilitation back home. (No refs.) - *E. L. Rowan.*

Charing Cross Hospital
Fulham, London S.W.6, England

3336 **MAC KEITH, RONALD.** Who guides the therapist? *Developmental Medicine and Child Neurology*, 13(1): 125-126, 1971. (Letter)

Improved care of children with speech problems could result from activity by pediatricians in promoting higher pay for speech therapists. Doctors working with neurologically handicapped children should understand those aspects of the disorder which are associated with speech handicaps and are amenable to the skills of the speech therapist. Since many speech therapists are newly qualified or inexperienced, guidance and encouragement are often needed. These therapists often have only a pediatrician available for consultation; consequently, knowledge by the latter of techniques of speech therapy, followed by appropriate communication, can be helpful. Speech therapists need the services of senior therapists for specialist assessment and suggestions for treatment. Difficult speech problems should be referrable to joint assessment clinics at regular intervals. (No refs.) - *M. S. Fish.*

No address

3337 **CONNOLLY, AUSTIN J.** Classroom forum. *Focus on Exceptional Children*, 3(3): 16, 1971.

Relevant topics and resources for in-service training in special education are noted. To develop an in-service training program, a school board and administration commitment should be obtained; a leader of the program should be acquired; administrative and instructional staff should explore potential goals for the program; commitments for financial support, time resources, and expertise should be secured; and an outline for a year's activities centered about a specific objective should be drawn up. (No refs.) - *T. C. Turner.*

University of Missouri
Columbia, Missouri

3338 **BUDNER, STANLEY; ARNOLD, IRENE; & GOODMAN, LAWRENCE.** The plan and the reality: Training and utilization

of paraprofessionals for service to the retarded. *American Journal of Public Health*, 61(2): 297-307, 1971.

A pilot program sponsored by Retarded Infants Services, Inc., has successfully trained low-income, unskilled personnel for ancillary service to professionals in MR. A total of 52 trainees in 3 groups was selected from over 275 applicants on the basis of an elementary school education, age over 18, former experience in working with children, motivation apart from experience with MR, and passage of a medical examination. The trainees were predominantly minority group members in the low-income bracket. The curriculum content of the program was based exclusively on future task requirements. Teaching methodology emphasized the concrete and practical, and the environment of the program was designed to afford as much status as possible. Eighty-seven per cent completed the 12 week training program, and 73% of these gained employment. The program had the additional effect of preparing individuals for employment in other health and welfare areas. (4 refs.) - *S. Glinsky.*

Fordham University School of Social Services
at Lincoln Center
New York, New York 10023

3339 **CYTRYN, LEON; & MILOWE, IRVIN D.** Development of a training program in mental retardation for psychiatric and pediatric residents. In: Menolascino, Frank J., ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation.* Seattle, Washington, Special Child Publications, 1971, 348-355.

A training program for psychiatrists, child psychiatrists and pediatricians in the framework of an MR institution has been developed. Evidence has indicated that most MRs cared for in residential institutions were admitted because of emotional disturbance, social maladaptation or family inadequacy. This substantiates the need for greater involvement of the psychiatric profession in MR. In the program residents interviewed MR children, held weekly psychotherapy sessions and interviewed parents. An increased sense of social responsibility was inculcated in the psychiatrists. Pediatric residents then participated and eventually fellows in child psychiatry entered the program. Of great benefit was the introduction of a didactic structure to the training program. Although problems remain to be resolved, the program successfully integrated the various specializations to the benefit of the MRs. (8 refs.) - *S. Glinsky.*

Children's Hospital Department of Psychiatry
Washington, D. C.

3340 **MENOLASCINO, FRANK J.** Mental retardation and comprehensive training in psychiatry. In: Menolascino, Frank J. ed. *Psychiatric Aspects of the Diagnosis and Treatment of Mental Retardation*, Seattle, Washington, Special Child Publication, 1971, 356-370.

Even if the psychiatric resident never sees an MR in his future practice, he will be a better practitioner if he has been exposed to MR in his training. A structured training in MR teaches the psychiatrist to think in terms of multifactorial etiological considerations, to assess intelligence better, to utilize psychopathology and psychodynamic factors, to program comprehensive treatment and habilitative services and to understand preventive aspects in MR. The result is a balanced, well-rounded psychobiological view of the personality which can be applied to any individual with complex or chronic problems. (35 refs.) - *S. Glinsky.*

University of Nebraska College of Medicine
Omaha, Nebraska

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